

# The Effect of COVID-19 Pandemic on Patients with Primary Immunodeficiency: A Cohort Study

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## What's Known

- As appropriate innate and adaptive immune responses are essential for SARS-CoV-2 eradication, some epidemiological studies have investigated the relationship between primary immunodeficiency diseases and the fatal risk of COVID-19.
- Recent studies suggests that patients with primary immunodeficiency in antiviral innate immune signaling or combined immunodeficiency may be predisposed to the severe form of COVID-19 infection.

## What's New

- Individuals with combined immunodeficiency are more susceptible to the severe form of COVID-19 infection than patients with other types of primary immunological deficiencies.
- Some primary immunodeficiency patients require more attention due to their age, category, and additional comorbidities.

## Abstract

Both adaptive and innate immune responses are essential for an effective defense against the severe acute respiratory syndrome Coronavirus 2 (SARS-CoV-2) infection. We aimed to investigate the effect of the coronavirus disease 2019 (COVID-19) pandemic on patients with primary immunodeficiency (PID). This study was performed on patients who were diagnosed with PID by immunologist specialists and referred to Imam Reza Clinic of Asthma and Allergy, affiliated with Shiraz University of Medical Sciences, (Shiraz, Iran) for regular check-ups. The patients were enrolled in this cohort study and followed for any sign of COVID-19 from March 2020 to May 2021. COVID-19 infection was confirmed using a real-time polymerase chain reaction (RT-PCR) assay of nasal and pharyngeal swabs. Among the 90 PID patients under study, nine patients (10%) were diagnosed positive for COVID-19 infection. Five out of these nine patients belonged to the combined immunodeficiency (CID) category, while four patients were categorized as having primary antibody deficiencies (PADs). Eight patients with COVID-19 were required to be admitted to the hospital, and three patients died after hospitalization due to COVID-19 infection. It seems that patients with CID are at a higher risk of mortality, due to COVID-19 infection, than other types of PID.

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**Keywords** • COVID-19 • Mortality • Primary immunodeficiency diseases

## Introduction

Currently, the world has encountered the coronavirus disease 2019 (COVID-19) pandemic caused by the severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2).<sup>1</sup> This virus can induce respiratory tract infection accompanied by cold, sneezes, pneumonia, and coughs.<sup>2</sup> Some conditions, such as obesity, old age, hypertension, male gender, cancer, diabetes, chronic lung disease, and heart conditions are considered as risk factors predisposing individuals to a severe form of the disease.<sup>3</sup> An orchestrated adaptive and innate immune responses is essential for an effective defense against SARS-CoV-2 infection without the negative side effects of a hyper-immune response.<sup>4</sup> In this regard, it is expected that patients with primary immunodeficiency (PID) may be more susceptible to developing the severe form of COVID-19.<sup>5</sup> Some studies have investigated

categories. These observations, along with other reports, either show that T cell response is more important against the virus or emphasize the role of B cells in the inflammation caused by SARS-CoV-2. Two out of the three PID patients died from COVID-19 infection, were under the age of three. In contrast with the results that show the mild form of the disease in children and adolescents to be due to the low expression of angiotensin-converting enzyme-2 (ACE-2) receptor and functional adaptive immunity, a fraction of them with PID who suffer from other comorbidities, may develop the severe course of the disease requiring hospital admission and even suffer a fatal outcome. In this regard, more caution should be exercised for complicated PID patients during the COVID-19 pandemic, and this should be taken into account by all physicians.

### Conclusion

In conclusion, it seems that patients with CID are at a higher risk of mortality, due to COVID-19 infection, than other types of PID. More attention should be paid to some PID patients regarding their age, the PID categories, especially CID, and further comorbidities. Future studies may confirm the individual risk of different PID diseases, and clarify the potential need for preventative measures for specific subsets of PID patients, who are at a high risk for a critical course of COVID-19.

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### Authors' Contribution

M.B: Conceptualization, Methodology, Formal analysis and investigation, original draft preparation, Supervision; Z.K: Formal analysis and investigation, original draft preparation, revise and editing; N.S: Formal analysis and investigation, original draft preparation; S.A: Conceptualization, revise and editing, funding acquisition, Resources, Supervision; All authors have read and approved the final manuscript and agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

**Conflict of Interest:** None declared.

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