Providing Enhanced Medical Support for Patients with Primary Immunodeficiency Disorders (PIDD): A Patient-Guided Intervention to Ensure Patient Engagement During a Global Pandemic and Beyond

Supported by an educational grant from Pfizer

The Survey

In 2021, the American Academy of Allergy, Asthma and Immunology (AAAAI) partnered with the Immune Deficiency Foundation (IDF) and AXDEV, a performance company specializing in evaluation and assessment, to solicit input from PIDD patients about the gaps they have experienced seeking care for immunodeficiency conditions. A Steering Committee made up of allergist/immunologists and patients associated with IDF drafted a survey addressing:

- Diagnosis
- Interprofessional collaboration
- Treatment and management
- Patient-provider communication
- Use of Telehealth

The survey was distributed by IDF to their community of PIDD patients, and over 300 patients submitted responses.

The Participants

<table>
<thead>
<tr>
<th>Adult Patients’ Diagnoses (n=308) PIDD patients over the age of 18</th>
<th>Age</th>
<th>Gender Identity</th>
</tr>
</thead>
<tbody>
<tr>
<td>56% (n=173) common variable immunodeficiency (CVID)</td>
<td>44% 61 or older</td>
<td>84% female</td>
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<tr>
<td>14% (n=44) hypogammaglobulinemia, IgG subclass deficiency or selective IgA deficiency</td>
<td>35% 46-60</td>
<td>16% male</td>
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<tr>
<td>17% (n=53) both of the above</td>
<td>27% 31-45</td>
<td>1% gender non-conforming</td>
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<tr>
<td>12% (n=38) other</td>
<td>3% 18-30</td>
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</tbody>
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<table>
<thead>
<tr>
<th>Provider Most Involved in Diagnosis</th>
<th>Provider Most Involved in Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>63% Immunologist or Allergist</td>
<td>80% Immunologist or Allergist</td>
</tr>
<tr>
<td>15% Other</td>
<td>8% Primary Care Physician</td>
</tr>
<tr>
<td>8% Pulmonologist</td>
<td>6% Hematologist</td>
</tr>
<tr>
<td>6% Infectious disease physician</td>
<td>4% Infectious Disease Physician</td>
</tr>
<tr>
<td>5% Primary Care Physician</td>
<td>2% Other</td>
</tr>
<tr>
<td>4% Hematologist</td>
<td></td>
</tr>
<tr>
<td>1% Mental Health Provider</td>
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</tbody>
</table>
The Findings

Diagnosis
Patients report long delays in diagnosis

- 41% (124/302) of adult sample reported that it took over 21 years beyond the onset of their first symptom to receive a final PIDD diagnosis
- 59% (179/304) reported that their PIDD was initially diagnosed as a different disorder (e.g., allergy, bacterial or viral infection, chronic sinusitis, Crohn’s disease)

Patients report the following potential reasons for long delays based on their experiences:

- HCPs do not ask patients about their full range of symptoms
  - 42% (126/302) of adult sample disagreed with the statement: When I initially consulted with a health care professional, they asked about the full range of symptoms I have been experiencing.

- HCPs tend to dismiss signs and symptoms of PIDD
  - 76% (226/296) agreed or strongly agreed with: I have experienced more than one healthcare professional completely dismissing my primary immunodeficiency related symptoms.
  - This is true even for patients who have common signs and symptoms of PIDD. Participants with the following symptoms agreed or strongly disagreed with this statement: “I have experienced more than one healthcare professional completely dismissing my primary immunodeficiency related symptoms.”
    - 91% Unexplained illness
    - 85% Recurrent pneumonia
    - 83% Chronic nasal or ear drainage
    - 81% Breathing problems
    - 80% Autoimmune disorders

Treatment and Management

- 91% (278/304) of adult patients reported that the severity of their symptoms worsened over time

Patient-Provider Communication

% of patients disagreeing or strongly disagreeing that HCPs explained to them how the following factors affect the severity of their PIDD

- 65% Environmental factors
- 60% Lifestyle
- 57% Another health condition

Telehealth

60% of adult patients reported perceived benefits associated with telehealth; some reported perceived limitations

- 25% provides little to no support for mental health
- 21% provides little to no opportunities for discussions on quality of life

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