Survey of Physician Attitudes and Practices Related to the Diagnosis and Management of Primary Immunodeficiency Diseases

Marking Instructions:
- Use a No. 2 pencil only rather than ink, ballpoint, or felt tip pens.
- Make solid marks that fill the response completely.
- Erase cleanly any marks you wish to change.
- Make no stray marks on this form.

1. What is your primary specialty?
   - Family Medicine
   - Family Medicine- Pediatrics
   - Family Medicine- Adult
   - Resident
   - Other, please specify below

2. In what type of ambulatory care setting do you spend MOST of your patient care time?
   - Solo practice
   - Single-specialty group practice
   - Multi-specialty group practice
   - HMO
   - Hospital outpatient
   - Other, please specify below

3. In an average week, about how many patients do you see on an outpatient basis? (Your best estimate is fine.)
   - patients per week

4. Have you ever followed patients with any of the following diagnoses? MARK AS MANY AS APPLY
   - Agammaglobulinemia (Bruton’s)
   - Ataxia Telangiectasia
   - Chronic Granulomatous Disease
   - Chronic Mucocutaneous Candidiasis
   - Common Variable Immunodeficiency
   - Complement Deficiency
   - DiGeorge Anomaly
   - Hyper IgM deficiency
   - IgA Deficiency (Selective IgA)
   - IgG Subclass Deficiency
   - Selective antibody deficiency to polysaccharide antigens
   - Severe Combined Immunodeficiency
   - Wiskott-Aldrich Syndrome
   - Any other primary immunodeficiency disease (SPECIFY)
   - None of these

5. Based on what you know or have heard, about what percent of patients with a primary immunodeficiency disease (PiDD) have a family history?
   - percent with family history

6. How many new patients do you expect to see in the next 12 months?
   - new patients expected in next 12 months

7. How many of those will have a primary immunodeficiency disease?

8a. Are you aware of any professional guidelines for the diagnosis and management of primary immunodeficiency diseases?
   - Yes
   - No skipping to Q9

8b. Who publishes those guidelines?

9. In what year did you graduate from medical school?
   - YEAR

10. How fully were primary immunodeficiency diseases covered in medical school?
   - Very well
   - Adequately
   - Only a little
   - Not at all

PLEASE DO NOT WRITE IN THIS AREA

[SERIAL]
11. What is your level of comfort with the recognition and diagnosis of primary immunodeficiency diseases?
- Completely Comfortable  PLEASE CONTINUE
- Very Comfortable  PLEASE CONTINUE
- Somewhat Comfortable  PLEASE CONTINUE
- Not At All Comfortable  SKIP TO Q35a

12a. Have you ever ordered tests or referred to a specialist a patient whom you suspected as having an underlying primary immunodeficiency disease?
- Yes, Ordered tests
- Yes, Referred to a specialist
- No, neither  SKIP TO Q14

12b. What tests would you NORMALLY order to establish a diagnosis of PIDD?  MARK AS MANY AS APPLY
- Antibody titers to vaccine
- CBC with manual differential
- CBC with automated differential
- CH50
- Quantitative serum immunoglobulins (IgG, IgA, IgM, IgE)
- IgG subclasses
- Neutrophil oxidative burst assay (DHR OR NBT)
- Chest x-ray
- Flow cytometry for lymphocyte subsets (T, B, NK cell subsets)
- Serum immunoelectrophoresis
- Blood tests for arterial gas levels
- CT scans
- Skin tests for allergic sensitization
- Anti-tetanus titer
- Anti-pneumococcal titer
- Anti-influenza titer
- DTH testing
- Other, please specify below

13a. Have you ever diagnosed a patient with a primary immunodeficiency?
- Yes
- No  SKIP TO Q14

13b. How many cases of PIDD have you diagnosed?

14. How effective is immunoglobulin replacement therapy (IgG therapy) in the treatment of antibody deficiency disorders?
- Very effective
- Somewhat effective
- Not too effective
- Not effective at all

15. Would you recommend IgG therapy for all, most, some, few or no patients with...

- Agammaglobulinemia
- Chronic Granulomatous Disease
- Common Variable Immunodeficiency
- Complement Deficiency
- DiGeorge Anomaly
- Hyper IgM
- IgA Deficiency
- IgG Subclass Deficiency
- Severe Combined Immunodeficiency

16. In counseling a PIDD patient considering IgG therapy, how would you present the risk of contracting the following diseases as a result of treatment?

- HIV
- Hepatitis B
- Hepatitis C
- CJD/Prion disease
- Rotavirus
- Yet to be discovered pathogens

No real risk
A small, but measurable risk
A moderate risk over time
A relatively high risk

17. For how many PIDD patients on IVIG would you recommend a surgically implanted line?
- All (91-100%)
- Most (51-90%)
- Some (11-50%)
- Only a few (1-10%)
- None (0%)

PLEASE CONTINUE TO NEXT PAGE
**18. Do you believe there is a greater than placebo effect benefit to any of the following alternative therapies for enhancing immunity, or preventing infection in patients with PIDD?** MARK AS MANY AS APPLY

- Massage therapy
- Acupuncture
- Vitamins
- Nutritional supplements
- Herbal formulations
- Probiotics
- Biofeedback
- Hypnotherapy
- Meditation
- Yoga
- Aerobic exercise

**19. Do you believe that any of the following hygiene related interventions offer a greater benefit than cost for patients with PIDD?** MARK AS MANY AS APPLY

- Alcohol-based hand gels for patient
- Alcohol-based hand gels for family
- Alcohol-based hand gels for classroom
- Regular soap and water hand washing
- Use of anti-bacterial soaps
- Avoidance of daycare
- Home schooling
- HEPA filter-based air purifiers in the home
- Dehumidification systems in the home
- Exclusion of furred pets from the home
- Exclusion of feathered pets from the home
- Use of disinfectant cleaners in the home
- Use of water filtration systems in the home

**20. For which PIDD patients do you recommend avoidance of live viral vaccination?** MARK AS MANY AS APPLY

- Agammaglobulinemia (Bruton's)
- Chronic Granulomatous Disease
- Common Variable Immunodeficiency (CVID)
- Complement Deficiency
- DiGeorge Anomaly
- Hyper IgM deficiency
- IgA Deficiency (Selective IgA)
- IgG Subclass Deficiency
- Selective antibody deficiency to polysaccharide antigens
- Severe Combined Immunodeficiency (SCID)
- Wiskott-Aldrich Syndrome
- Other (SPECIFY)
- Would not recommend avoidance for any

**21. For how many patients are you the primary treatment provider for their PIDD, and for how many patients do you follow but are not the primary treatment provider for their PIDD?**

<table>
<thead>
<tr>
<th>Primary treatment provider for PIDD</th>
<th># of patients</th>
<th>Follow patient, but not primary for PIDD</th>
</tr>
</thead>
<tbody>
<tr>
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</tbody>
</table>

**22a. What proportion of the PIDD patients you follow do you co-manage with a sub-specialist?**

- All
- Most
- Some
- None

**22b. Which type of sub-specialist do you most commonly work with for PIDD patients?**

- Allergy
- Allergy/Immunology
- Hematology
- Immunology
- Other medical professionals (SPECIFY)

**23. Do you use prophylactic antibiotic therapy for your patients with PIDD?**

- Yes
- No

**24. About how many PIDD patients do you follow who are maintained on IgG therapy to prevent infections?**

<table>
<thead>
<tr>
<th>PIDD patients on IgG therapy</th>
</tr>
</thead>
<tbody>
<tr>
<td>None</td>
</tr>
</tbody>
</table>

**25. What percentage of your PIDD patients receive their IgG therapy at the following sites --- all or almost all, most, some, few to none?**

<table>
<thead>
<tr>
<th>Few to None (&lt;5%)</th>
<th>Some (5-50%)</th>
<th>Most (&gt;50%)</th>
<th>All or almost all (&gt;95%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Your office</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Hospital outpatient infusion suite.</td>
<td></td>
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<tr>
<td>Hospital inpatient</td>
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<tr>
<td>Home with a nurse.</td>
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<td></td>
<td></td>
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<tr>
<td>Home via self-infusion.</td>
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</table>

**PLEASE CONTINUE TO NEXT PAGE**
26. Do you manage the IgG therapy for any patient with PIDD?
- Yes
- No  SKIP TO Q30

27. What is the usual interval with which you recommend patients with antibody disorders receive their IgG therapy?
- 1 week or more often
- 2 weeks
- 3 weeks
- 4 weeks
- 5 weeks
- 6 weeks
- More than 6 weeks

28. What is the usual IgG therapy dosage per kilogram of body weight that you typically recommend for patients with antibody disorders?
- < 200 mg per kg
- 300 mg per kg
- 400 mg per kg
- 500 mg per kg
- 600 mg per kg
- > 600 mg per kg

29. In the past 12 months, have you had problems with reimbursement for IgG therapy for PIDD patients covered by...? MARK ALL THAT APPLY
- Medicare
- Medicaid
- Private insurance
- None of these

30. How much risk do current reimbursement standards for IgG therapy pose to the health of PIDD patients?
- Extreme risk
- Serious risk
- Moderate risk
- Slight risk
- No real risk

31a. How familiar are you with subcutaneous immune globulin replacement therapy?
- Very familiar
- Somewhat familiar
- Not too familiar

31b. Do you use subcutaneous immune globulin replacement therapy for PIDD patients?
- Yes
- No

32. Approximately, what proportion of patients and families you’ve treated with mild to moderate PIDD are satisfied with the management of their disease?
- All (91-100%)
- Most (51-90%)
- Some (11-50%)
- Few (1-10%)
- None (0%)

33. What proportion of patients and families you’ve treated with PIDD have received education about their disease and prognosis?
- All (91-100%)
- Most (51-90%)
- Some (11-50%)
- Few (1-10%)
- None (0%)

34. What proportion of patients and families you’ve treated with PIDD understand their disease and prognosis?
- All (91-100%)
- Most (51-90%)
- Some (11-50%)
- Few (1-10%)
- None (0%)

35a. Have you heard a lecture regarding PIDD recognition and diagnosis in the last 6 months?
- Yes  SKIP TO END OF SURVEY
- No

35b. Do you feel this would be beneficial to your patients?
- Yes
- No

PLEASE PROVIDE YOUR NAME AND ADDRESS SO WE MAY SEND YOU THE CHECK:
Name:______________________________________
Address:______________________________________
City:__________________________________________
State:_________________________ ZIP:____________

Please return the questionnaire in the enclosed envelope to: Immune Deficiency Foundation
40 W Chesapeake Avenue, Suite 308   Towson, MD 21204

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