

Indian Academy of Pediatrics (IAP)



## GUIDELINES FOR PARENTS

# Care of a Child with Immune Deficiency

*Convener:* **Anju Gupta**

*Members:* **Ambreen Pandrowala,  
Baldev S Prajapati,  
Rupam Das**

*Reviewer:* **S Sitaraman**



### 11 FAQs on CARE OF A CHILD WITH IMMUNE DEFICIENCY

1. What is immune deficiency? What can cause it? Can these diseases be passed on to our next child, if one child is already affected? What can be done to prevent it?
2. My child keeps on getting infections every few days. Should I worry about immune deficiency?
3. What infections can my child get? When should I take him/her to a hospital? What can I do as a parent to prevent infections?
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8. What is the role of homeopathic/complementary/alternative therapies in treatment of primary immune deficiency? Is there any benefit of giving immune boosters to my child with immune deficiency?
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10. Are there any patient advocacy groups in India for children with immune deficiencies?
11. What are the common types of immunodeficiencies in children?

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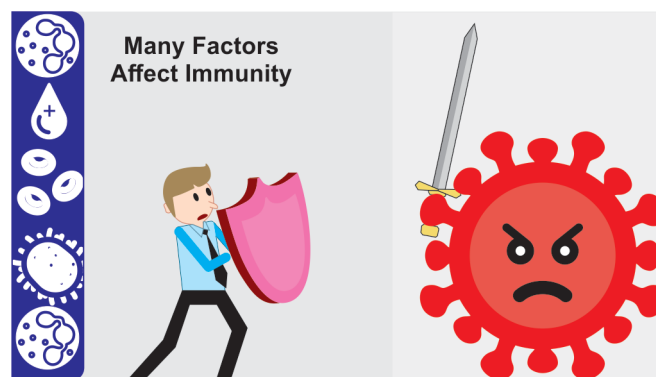
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# Care of a Child with Immune Deficiency

## Q1

**What is immune deficiency? What can cause it?  
Can these diseases be passed on to our next  
child, if one child is already affected? What can  
be done to prevent it?**

The immune system helps to protect us from infections. Immune system has many components, and different components are important for protection against different bugs (**Fig. 1**).



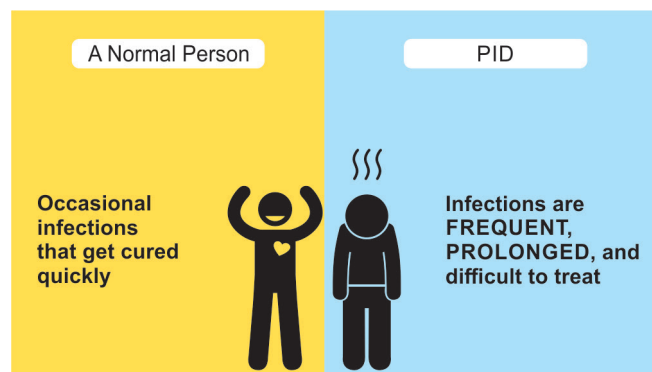
**Fig. 1:** Immune system protects us from different bugs.

When any component of this system fails, we call the condition an “immunodeficiency” (**Fig. 2**). Primary immune deficiency (PID)

is not a single disease. Based on which component of immune system has failed, PID can be of many types. Presently, >450 different diseases are grouped under PID. They differ in their manifestations, treatment, and outcome. But, core theme of all these diseases is frequent and prolonged infections (**Fig. 3**).



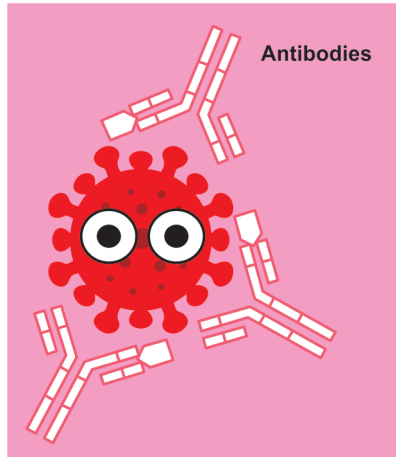
**Fig. 2:** Failure of any one component of immune system can cause primary immune deficiency (PID).



**Fig. 3:** Patients with primary immunodeficiency tend to have frequent and prolonged infections.

Outcome of PID is also different depending on type of PID. Some immunodeficiencies are severe and can cause symptoms within first few months of birth such as severe combined immunodeficiency (SCID). Most of the children with SCID die during first year of life unless treated appropriately. Some of the PID are less serious and may even be diagnosed in adults.

Antibody deficiency is the largest group of PID worldwide. In this group of PID, children do not form certain proteins called "antibodies". These antibodies help in killing the bugs and, hence, protect us from infections (**Fig. 4**). In the absence of these antibodies, these children catch frequent infections.



**Fig. 4:** Antibodies help us in killing the bugs and protect us from infections.

The cause of PID is usually due to a faulty gene. The fault may be a new change in one of your child's genes, or it may have been passed on from one or both of the parents. Most of PID cannot be cured, but with proper care, most children and adults can enjoy a reasonable quality of life.

Because the cause of PID lies in faulty genes, these diseases can be passed on from one generation to the next in the family. The chances of the next child getting PID vary depending on type of PID. Testing to determine whether unborn babies are affected is possible at many centers in India. Please speak to your doctor about the same.

## Q2

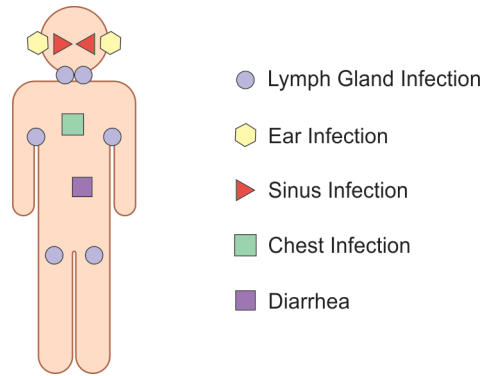
### My child keeps on getting infections every few days. Should I worry about immune deficiency?

It depends on type of infections. It is very common to have up to 10–12 respiratory infections presenting as fever, cough, sneezing, and runny nose every year in children. Such infections do not warrant investigations for immune deficiency, if your child has normal growth and development, recovers completely from such infections within 5–7 days, and appears healthy in-between infections.

Children with PID tend to develop chest infections, ear and sinus infections, diarrhea, and infections of lymph glands (**Fig. 5**).

The Jeffrey Modell Foundation has suggested 10 warning signs for immunodeficiency (**Fig. 6**) (<http://downloads.info4pi.org/pdfs/10-Warning-Signs---Illustrated--2-.pdf>). These warning signs include:

- Four or more new ear infections within 1 year
- Two or more serious sinus infections within 1 year
- Two or more months on antibiotics with little effect
- Two or more pneumonias within 1 year
- Failure of an infant to gain weight or grow normally
- Recurrent, deep skin or organ abscesses
- Persistent thrush in mouth or fungal infection on skin
- Need for intravenous antibiotics to clear infections
- Two or more deep-seated infections including septicemia
- A family history of PID



**Fig. 5:** Common sites of infections in children with primary immune deficiency (PID).

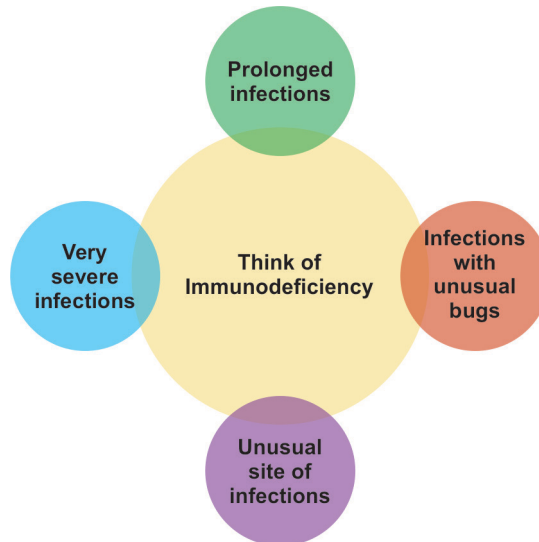
### Warning Signs for Primary Immunodeficiency



**Fig. 6:** Warning signs for primary immunodeficiency developed by the Jeffrey Modell Foundation.

*(These warning signs were developed by the Jeffrey Modell Foundation Medical Advisory Board. Consultation with Primary Immunodeficiency Experts is strongly suggested)  
Source: Reproduced with permission from ©2016 Jeffrey Modell Foundation.*

If your child has two or more of these warning signs, you should definitely talk to your doctor regarding a possibility of PID. Infections with unusual bugs, unusual severity, unusual sites and/or unusual duration raise the concerns about PID (**Fig. 7**).



**Fig. 7:** Infections in primary immune deficiency (PID).

While infections are the most common presentation of PID, other features such as joint pain and swelling, skin rash, and destruction of red blood cells (anemia) can also be present. These features occur when the body attacks against its own cells, which is called “autoimmunity”.

**Q3**

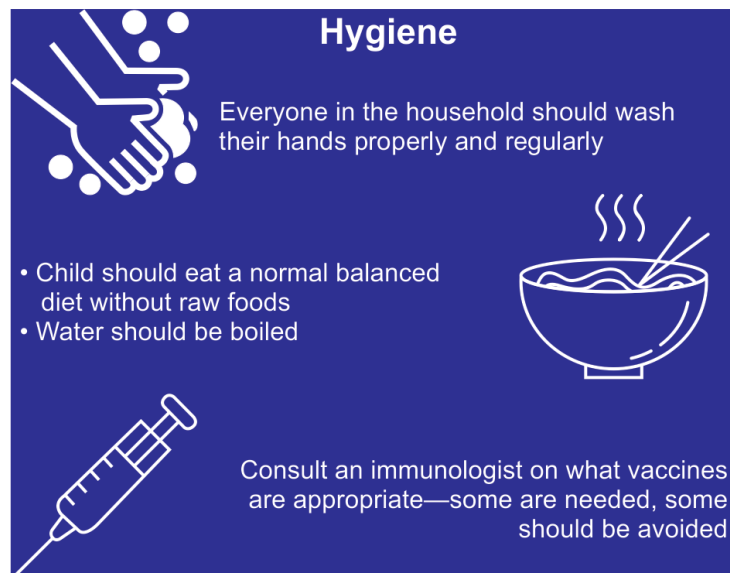
**What infections can my child get? When should I take him/her to a hospital? What can I do as a parent to prevent infections?**

Parents of children with PID need to be vigilant and must contact their doctor if the child has symptoms of infection such as fever, throat pain, swellings at any site, lethargy, irritability, reduced appetite, diarrhea, and vomiting. Other symptoms may depend on the body part affected by the infection. All infections do not need hospital admission. Your doctor will assess whether the infection can be treated at home or needs hospital admission.

Keep all medical records in a safe and easily accessible place in case the child needs hospitalization. The school authorities should also be informed about the medical condition of the child.

As a parent, it is important to follow and teach your child good hygiene practices to prevent infections. Here are some tips for good hygiene (**Fig. 8**):

- Handwashing before meals, after outdoor activities, after playing with pets, and after using the toilet should be a routine. Hands must be washed vigorously with soap and water for at least 30 seconds.
- If handwashing is not feasible, then use alcohol-based hand rubs.
- Encourage family and friends to follow strict hand hygiene so they do not spread infections to the child.
- Wash all cuts with soap and water, and then apply antiseptic.
- Keep immunizations up-to-date as recommended by your immunology team.
- While good dietary habits are important for everyone, they are extremely important for children with PID. Children, in particular, need a balanced diet to grow and develop normally. Practice good food hygiene to avoid food poisoning.
- Water needs to be boiled before drinking. Boiled water can be stored in the refrigerator or at room temperature for 24 hours, if the container being used to store the water has a lid.
- Children with PID can eat usual foods. It is better to take more precautions against germs because they are more vulnerable to infections. Avoid raw or undercooked dishes.
- Wash all toys regularly.



**Fig. 8:** Useful hygiene tips for a child with primary immune deficiency (PID).



Q4

### Can I send my child with primary immune deficiency (PID) to school?

Children with PID should be sent to school. It is important for their self-confidence (**Fig. 9**). School absenteeism can be an issue due to frequent hospital visits for follow-up and sometimes due to infections. Despite this, children with PID can do very well in studies with a little extra support from parents and school.

It is important for parents to meet with teachers, counselors, and administrators to explain their child’s condition and its potential impact such as frequent absences, fatigue, and activity restriction.

**Your child with careful precautions can do many things**

- School can help!
- Keep teachers informed about your circumstances



 Your doctor should be consulted before holidays. Take care of food hygiene.

Sports are encouraged, but avoid swimming, gardening and contact sports depending on the PID



**Fig. 9:** Children with primary immune deficiency (PID) can do a lot with some precautions.

Good hygiene practices should be followed in the school. Frequent handwashing and respiratory hygiene should be followed as well. It is better to stay away from an obviously infected fellow student.

Q5

### Can my child play outdoors?

- Children with PIDs can enjoy exercises and sports, just like everyone else, and they should be encouraged to do so.
- Children with certain PID such as chronic granulomatous disease (CGD) should avoid swimming in lakes or ponds, and gardening.
- Certain PIDs interfere with blood clotting and such patients should avoid contact sports.

Q6

### Can we take our child on a holiday? Should we take any precautions?

Children with PID can travel on a holiday with some extra precautions. If the child is traveling abroad, then same should be discussed with the doctor. Travel to some countries requires vaccination for certain diseases prior to travel. Your doctor will be able to inform you if the required vaccines are safe for your child. If the vaccine is not safe, your doctor will issue an exemption certificate. During travel, food hygiene should be taken care of. It is important to avoid unboiled water including ice cubes. Parents should know how to contact local health system, in case the child develops any symptoms during travel.

#### Few Important Tips

- Medication and supplies should be packed in a way that they are easily accessible during travel.
- Keep your medicine in the original container with proper prescription.
- Drink boiled water.
- Make sure that meat, poultry, shellfish, and fish are completely cooked.
- Fruits and vegetables should be washed and peeled.
- Keep contact details of your child's immunologist.
- Always carry a copy of your insurance card.

Q7

### **Should my child's vaccination be different from vaccination of children with no immune deficiency?**

Your child may need additional vaccines or may need to avoid certain vaccines. Your specialist doctor will discuss this with you and advise on which vaccines are safe for your child.

The families of children with PIDs should normally be vaccinated in order to protect these children from catching infections including the influenza vaccine. Your doctor will guide you regarding it.

There are certain restrictions for vaccination, which your doctor will discuss with you. If you have had a previous child with diagnosed PID, and you have another baby, please do not get the baby vaccinated at birth and contact your doctor immediately.

Q8

### **What is the role of homeopathic/ complementary/alternative therapies in treatment of primary immune deficiency? Is there any benefit of giving immune boosters to my child with immune deficiency?**

Homeopathic, complementary, and alternative therapists claim that they can cure or improve immunity by medicines. But, there is no valid scientific data or research to prove that claim, so it is difficult to comment regarding the efficacy and safety of alternative medicines.

No immune boosters can help to replace parts of immune system that are not working. Good nutrition, appropriate exercises, and good sleep are beneficial.

## Q9

**What treatment options are available in India for a child with immune deficiency?  
Where can I get these facilities?****Treatment of PID Depends on Type of PID**

The largest group of PID worldwide is “antibody deficiency”. Antibodies are special proteins in our body, which help in protecting us from infections. Replacement of these antibodies helps these patients by keeping them free of infections. This is called “immunoglobulin replacement therapy”. Immunoglobulin replacement is given intravenously once a month. Some of these patients are also given small dose of an antibiotic daily to prevent infection. If even after these measures, these children develop infections, the infections need to be treated early and aggressively.

In some other type of PID, patients are given antibiotic and/or antifungal medicines daily to prevent infections. More specialized treatment for some types of PID includes hematopoietic stem cell transplantation. Stem cells are “master cells”, which produce various cells of the immune system. The transplant aims to correct the immune defect by replacing the diseased stem cells with healthy stem cells. Hematopoietic stem cell transplant is not useful in all forms of PIDs. Talk to your immunologist, if transplant is recommended for your child’s disorder.

Immunology clinics are available in India at many centers. Updated list of these centers is available at <https://fpid.org/wp/fpid-centers-in-india/>. Further details regarding PID centers may also be found at website of Indian Society for Primary Immune Deficiency (ISPID) <http://ispid.org.in/> which works in close liaison with Foundation for Primary Immunodeficiency Diseases (FPID), USA.

## Q10

### Are there any patient advocacy groups in India for children with immune deficiencies?

Primary immune deficiencies are rare and there is lack of awareness about these disorders among physicians and public in India. An online story for parents and children with PID is available at <http://pinsa.org.za/wp-content/uploads/2014/10/Kids-Our-Immune-System.pdf>.

There are a few patient advocacy groups in India, which work for families of children with PID.

- *Indian Patient Society for PID (IPSPI)*: Based in Bhubaneswar. <https://www.ipspiindia.org/>
- *Primary Immune Deficiency Patients Welfare Society (PIDPWS)*: Based in Bengaluru.
- The International Patient Organization for Primary Immunodeficiencies (IPOPI) is a patient advocacy group for children with PID with worldwide presence including India.

These organizations work to improve the quality-of-life of individuals with PID in India.

## Q11

### What are the common types of immunodeficiencies in children?

As discussed before, >450 PIDs are known today. Fortunately, a few disorders are responsible for majority of PIDs seen in the population. We will discuss a few common disorders in brief:

- *Antibody deficiency disorders* are the most common group of PID worldwide. Antibodies (immunoglobulins) are proteins made by our body and they help us in fighting infections. Typical antibody deficiency disorder seen in childhood is X-linked agammaglobulinemia. In this disorder, the body cannot make antibodies due to a genetic defect. These children develop frequent ear, sinus, and chest infections. Most of these children would have received multiple courses of antibiotics for infections before

a diagnosis is made. Replacement of antibodies is done by giving monthly injections of immunoglobulins. This treatment is expensive and needs to be given lifelong. Early diagnosis is important for a good outcome, because repeated chest infections can cause permanent lung damage.

- *Severe combined immunodeficiency (SCID)* is a group of serious immunodeficiency disorders that causes death within first year of life. These babies develop infections with unusual bugs and do not clear infections even with appropriate treatment. Early hematopoietic stem-cell transplantation can save life.
- *Chronic granulomatous disease* is another group of immunodeficiency disorders in which certain cells responsible for killing bugs are defective. These children can develop infections at any age. Infections with unusual bugs are common. Common infections involve chest, skin and lymph glands in neck, axilla, and groin. Some of these children do well on lifelong medicines to prevent infections, whereas others need early hematopoietic stem-cell transplantation.
- Wiskott–Aldrich syndrome is a serious immunodeficiency disorder seen in boys. These children present during first year of life with skin bleeding and bloody stools. Skin eczema is common. These children develop frequent infections and autoimmunity. Hematopoietic stem-cell transplantation is the only known cure of this disorder.

### KEY POINTS

- Our immune system is the main defense of our body against infections. It has many components and failure of any component can cause immune deficiency.
- Many of the immune deficiencies are genetic and can be passed on from one generation to the next. Main presentation of PID is frequent and prolonged infections.
- Some of the PIDs are very severe and cause death within first year of life, if not treated appropriately. Other PIDs may be milder and may even present in adulthood.
- Good diet, appropriate exercises, good sleep, and good personal hygiene are important for every child with PID besides appropriate and early treatment of infections. Specific treatment differs depending on type of PID.