



IPOPI

INTERNATIONAL
PATIENT ORGANISATION
FOR PRIMARY IMMUNODEFICIENCIES

PRIMARY IMMUNODEFICIENCIES

PRIMARY IMMUNODEFICIENCIES AND **PSYCHOLOGICAL MANAGEMENT**



INTRODUCTION

This booklet discusses the psychological effects that primary immunodeficiencies (PIDs) can have on patients, parents, carers and families, and suggests ways to help everyone concerned to cope with these effects.

Primary immunodeficiencies (PIDs) are rare, chronic, inherited diseases that occur when components of the immune system are either not present or are not functioning normally. By interfering with this system, PIDs leave people susceptible to infections. There are many different types of PID, ranging from relatively mild conditions to life-threatening diseases. Some PIDs increase the risk of cancer, while others can cause 'auto-immune' disease whereby the immune system attacks parts of the patient's own body (e.g. the joints, causing arthritis).

The prognosis of PIDs has improved greatly with the use of immunoglobulin replacement therapy and other modern therapies. However, as well as their effects on the body, PIDs can profoundly affect patients' psychological wellbeing, quality of life and mental health, and can cause stress, anxiety and depression. This impact extends to parents, partners and other family members, who play a vital role in the care and support of patients with PIDs.

PIDs affect people in many different ways. Episodes of severe, acute illness can be very distressing, while in the longer term the unpredictable future effects of PIDs can create uncertainty and fear throughout life. People affected by PIDs can sometimes feel they lack control over their lives, and this can cause feelings of hopelessness and despair. Because PIDs are chronic and rare, patients may feel that the people around them do not understand their concerns and as a result they may feel isolated and unsupported. The medical care that patients need can itself have psychological effects, e.g. via the disruption and anxiety caused by clinic visits, medical tests and hospital admissions (e.g. for infections) and concerns about the side effects of long-term treatments.

This booklet looks in more detail at the psychological impact that PIDs can have, specifically in children and their parents, and in adolescent and adult patients. Importantly, it also suggests various things patients and everyone affected by PIDs can do to help cope with these effects, and highlights many sources of support.

KEY ABBREVIATIONS

CVID	Common Variable Immunodeficiency
IPOPI	International Patient Organisation for Primary Immunodeficiencies
PID	Primary immunodeficiency

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CHILDREN AND PARENTS

ISSUES SPECIFIC TO CHILDREN

PIDs can affect the psychological and social development of children in many ways. Children may have problems understanding their disease and why they have it. They may also be frightened of what might happen to them, or of the medical examinations, procedures and treatments they need. Indeed, children may be reluctant or unwilling to take regular medication, which can cause family conflict. In turn, parents may find it difficult to talk to a child about their PID and to explain about the necessary tests and treatments.

Absences from school due to illness can interfere with the education of children with PIDs. PIDs can also limit childrens' social interaction with other children and their participation in school activities (such as some sports) and leisure pursuits (for example, those that carry a risk of infection). Children may be distressed or angry if they have to miss out on these activities. They may feel lonely and embarrassed owing to feelings of being 'different' to their peers, and they may even face marginalisation or bullying. It is important to recognise that children, just like adults, can suffer depression and anxiety.

The siblings of children with PIDs can also be affected in many ways by their sibling's disease. Siblings can feel upset, anxious, or confused about their brother or sister's health. They may also feel excluded or jealous about the attention that their sibling receives because of his or her PID.

PARENTS

A diagnosis of a PID can affect the wellbeing of parents in many ways. The complications of severe PIDs, such as life-threatening infections, can be highly distressing. In the longer-term, parents at times can feel sadness, despair and hopelessness in facing their child's chronic illness. In some cases, parents may have difficulty in coming to terms with the reality of their child's disease. In some areas of the world, they may sometimes feel overwhelmed and desperate because they do not have medical insurance coverage, or sufficient funds, to deal with their child's treatment. Since PIDs are inherited, genetic diseases, parents commonly experience feelings of self-blame and can even face animosity from other family members and resentment from affected children. These challenges can be exacerbated by stress caused by the financial difficulties that can occur if a parent needs to take time off work, or even give up work, to care for a child with a PID.

WHAT CAN BE DONE TO HELP?

The diagnosis of a PID in a child affects the whole family and measures to cope with its impact should involve everyone.

Parents are encouraged to aim to create as normal a life as possible for a child with a PID and the rest of the family. The extent to which this is possible, and the potential ways to achieve it, will differ for each family and depend on the circumstances. Generally speaking, it involves thinking positively and creatively to find ways to allow home life, school and social activities, and other choices that enrich life while realistically balancing these with the restrictions of the PID. PID patient organisations can provide advice, examples and support from other parents who have faced similar situations.

TESTIMONIALS

- "I think it is very important to share experiences and I know from many conversations with other members of my PID patient organisation that it is important to know each other and to network because in your social circle you usually do not have other PID patients. Members can exchange day-to-day worries, like what to do in kindergarten, what is the best way to send the child to school, etc. – all the things a doctor cannot answer." Kerstin, mother of a patient with agammaglobulinemia and member of the dsai PID organisation in Germany
- "PIDs always impact the lives of patients, but it is necessary to prevent that impact from being strong. In the case of my son, we try to make that impact as minor as possible. We make all efforts to make his life as normal as possible in his studies and we are very happy. This is a work that starts at birth and it is a continuous effort. The better you take care of yourself the less the impact will be in your daily life." Carlos, father of a PID patient in Spain

More testimonials from parents can be found at www.tv.ipopi.org.

Practical ways to help the whole family cope with a PID include:

Communication within the family: Parents and carers should make sure to obtain accurate, up-to-date information on their child's PID from reliable sources. Resources and support are available to help parents talk with a child (and their siblings) about their PID, to give clear, realistic and age-appropriate information, and to answer their questions. It is important that parents communicate with each other and with the wider family.

School: Most children with PIDs can attend nursery and school and educational trips normally. A companion IPOPI booklet (*Primary Immunodeficiencies: a guide for schools*) explains how schools can best support students with PIDs. Parents are encouraged to seek information about their rights and the local procedures through which they can obtain the best support and opportunities for their child with a PID and to discuss their child's situation and needs with school staff.

Activity: Most patients with PIDs can partake in most activities, including sports. Indeed, exercise is just as important for both the mental and physical health of children with PIDs as for everyone else. Where limitations are necessary, parents may be able to work with schools and clubs to find ways to help children with PIDs participate and integrate as far as possible.

Treatment: Careful scheduling of appointments may help to lessen the impact of treatment on schooling and family life. In some circumstances, immunoglobulin therapy can be given at home, which can be more convenient for patients, families and carers. The IPOPI booklet, *'One size doesn't fit all'* provides further information on home-based therapy.

Counselling: Some parents and children may benefit from psychological counselling or stress management to help them deal with their disease. It is important that parents explain the emotional effects of their child's illness to the doctor or nurse and discuss available support. Children with PIDs should also be encouraged to talk about these issues. Parents who lose a child to a PID may benefit from grief counselling.

ADOLESCENTS WITH PIDs

The emotional and hormonal changes that occur during adolescence add particular challenges for young people with PIDs. Adolescents may experience emotional difficulties associated with relationships, home life and the pressures of education and work, all of which can compound the impact of their PID. Adolescents are also faced with the need to take more responsibility for their self-care and for a transition between children's and adult healthcare services, where these are separate. Some adolescent patients may have difficulty taking on these roles, while some parents may find it difficult to hand over these responsibilities to their child.

During this point in life, PIDs can exacerbate an adolescent's feelings of abandonment or poor self-esteem. Feelings of resentment at having inherited a PID may surface. Some adolescents may feel disappointed or even angry about the limitations that their PID imposes on employment, training, study, travel or other life choices.

WHAT CAN BE DONE TO HELP?

Most of the measures discussed on page 5 are also important for adolescents, but they need to be tailored to situations faced by this group. We encourage young people, and everyone around them, to take advantage of all available sources of information and support to help balance a young person's opportunities with the restrictions associated with the PID.

Communication and motivation: It is very important that adolescents can communicate their feelings to the people around them. Contact with peers and role models who have PIDs may be helpful in providing emotional support, advice and motivation. Counselling may be helpful in some situations. Young people should be encouraged to seek such support without feelings of stigma.

Self-care education: Adolescents need to have access to clear, accurate and up-to-date education about their self-care to allow them to take on a greater role in day-to-day self-care and in making decisions about their treatment. Young people with PIDs need to understand about the symptoms of infections and other complications that may warrant medical attention, and how to get medical attention if they need it. Young people might benefit specifically from information and advice sources about sexual health and relationships' issues.

Home-based treatment: Many people who use immunoglobulin therapy can self-treat themselves at home. This can be more convenient and less disruptive than clinic-based treatment, although patients need to learn the necessary methods and it is not suitable for everyone (see the *'One size doesn't fit all'* IPOPI booklet).

Transition planning: Careful planning, and an individualised approach, can help to minimize the disruption that can occur when young people transition between paediatric and adult services. A companion booklet *'Moving from child to adult care'* explains these issues in more detail.

Education and careers advice: Having a PID should not prevent most patients from getting a full education or doing the sort of job they would like to do. It may help to consult school, college or university liaison staff to discuss any necessary support. Careers' advice services may be able to help patients to understand and explore the opportunities that exist for training and employment.

TESTIMONIALS

- “I started intravenous immunoglobulin at the age of 13 in Hong Kong. At the age of 17, when I moved to the UK to study, the doctor recommended I try subcutaneous immunoglobulin. My health condition improved. When I finished my A levels I returned to Hong Kong to continue my studies at university and was put back on intravenous immunoglobulin.” Geoff, a PID patient from Hong Kong
- “My disease affected me socially a lot more in the past than it does presently. I isolated myself a lot, I wouldn’t let people get close nor would I approach them. I used to hide behind my books! But as I turned 15 years old, it became easier for me to socialise. Those around me were more empathetic in the sense that they would show some interest in knowing how I was doing. There was an openness in mentality from my side, and also from those around me. Talking about my PID made me feel better in my own skin.” Rita, a Portuguese patient with COVID.
- “I think transition should start very early. We thought that the most important mission for us as parents was to give our son the tools to be able to live an independent life. Already at the age of 6, when we were at the hospital, he started to meet his doctor for a few minutes alone. At the age of 13 he started to go to hospital visits by himself. Step-by-step he has learned a lot about his own disease and how to take care of himself.” Anneli, mother of a patient with PID in Sweden.

ADULTS WITH PIDs

SPECIFIC ISSUES

People with PIDs are living longer than ever because of improved treatment. While this is a good thing, it also means that more and more people must learn to cope with the long-term psychological effects of having a chronic PID. Many people might be afraid to talk about these sorts of problems owing to stigma sometimes placed on mental health issues.

PIDs can cause particular stress on patients and families when they affect the ability to work or result in frequent work absences (for example due to infections or regular appointments) and hence cause financial difficulties. Sometimes PID patients might continue to work despite feeling unwell, which can affect their health as well as their performance at work. Others eventually need to retire early because of their PID.



Adults with PIDs also face difficult decisions associated with family planning, in light of the possibility of genetic inheritance. These considerations can include whether to have a child, whether to use genetic tests to assess the risk of the PID in an embryo or foetus, or whether to use donor eggs or sperm via in vitro fertilisation (IVF).

As many more people with PIDs reach older age, they face the stress of limited mobility and reduced independence. They may also worry about age-related complications of their PID (such as cancer, for some PIDs) and the onset of other age-related health problems. Older people can often feel isolated, leading to problems of loneliness and depression.

WHAT CAN BE DONE TO HELP?

The support of friends and family remains important to people with PIDs at all ages. As the community of adults and elderly people with PIDs grows, individuals can also expect to find more and more peers who can offer advice and support, for example through PID patient organisations. Patients may also benefit from contact with communities with other types of chronic and genetic diseases that create similar problems, and with mental health support groups.

It is important to continue to explore ways to help maintain good physical and mental health, and a sense of control, throughout life. For example, adults are encouraged to find opportunities to exercise and stay active regardless of their age. Home-based self-treatment, where appropriate, can help to improve patients' wellbeing by helping them to be more independent and to travel. Patients are encouraged to stay up-to-date with treatment options and to discuss these with their doctor.

Patients are also encouraged to seek out and take advantage of other local or national support and advice services that may be available. For example, these can include:

- Family planning and genetics counselling services, to discuss the implications of the PID and the availability, benefits and risks of different tests and options.
- Occupational health services, to help patients to maintain the ability to work.
- Employment advice services, regarding employment rights and opportunities for changing career
- Social welfare, support and advice services, to help patients understand and navigate the services that are available locally.
- Some patients may benefit from counselling or even medical therapy for mental health problems and are encouraged to discuss the need for this with their doctor without fear of stigma.

TESTIMONIALS

"I was 45 years old when I was diagnosed. Getting diagnosed was very important for me. Before I was diagnosed my quality of life was very poor. I missed lots of school days and later, at work, I ran the risk of losing my job. Because of my subcutaneous immunoglobulin treatment my work absence has decreased and my quality of life has increased. This is the most significant difference. I thank all of those involved in making my life the way it is now – just fine." Lutz, a patient with CVID from Germany

WHO CAN I ASK FOR HELP?

SUPPORT NETWORKS AND PATIENT ORGANISATIONS

PIDs affect the family and support networks of patients, as well as patients themselves. Family members, friends, schools, colleges, workplaces, together with healthcare and support services' staff, can all play very important roles in helping patients to cope with PIDs.

Although patients and families affected by PIDs may feel isolated at times, they are never really alone. There are many people out there who understand the problems that PIDs cause and who can help by offering contact, support, advice and a community spirit. Numerous patient groups and networks provide information, discussion forums and information via websites and social media and at events. Please contact IPOPI to get in touch with the patient group closest to you. IPOPI's own website contains testimonials by patients and parents (see www.tv.ipopi.org), as well as information, news and coverage of events.

PROFESSIONAL HELP

At times many patients, parents and carers need the help of a healthcare professional, such as a counsellor or psychologist. Signs that professional help may be needed include persistent low mood or sadness, feelings of hopelessness, anxiety and the feeling of being overwhelmed, problems sleeping and difficulties in working or in home, family and work life. Patients, carers and families affected by these sorts of problems should contact their doctor to discuss how to manage these effects and the support services that are available, such as a psychologist, counsellor and stress management. Alternatively, patients, parents and carers may also be able to obtain these services on a private basis.

FURTHER INFORMATION AND SUPPORT

This booklet has been produced by the International Patient Organisation for Primary Immunodeficiencies (IPOPI). Other booklets are available in this series. For further information and details of PID patient organisations in 63 countries worldwide, please visit www.ipopi.org.

Provided by



Supporting families affected by primary and secondary immunodeficiency

Immunodeficiency UK is a national organisation supporting individuals and families affected by primary and secondary immunodeficiency.

We are the UK national member of IPOPI, an association of national patient organisations dedicated to improving awareness, access to early diagnosis and optimal treatments for PID patients worldwide.

Our website has useful information on a range of conditions and topics, and explains the work we do to ensure the voice of patients with primary and secondary immunodeficiency is heard. If we can be of any help, please email us or call on the number above, where you can leave a message.

Support us by becoming a member of Immunodeficiency UK. It's free and easy to do via our website. Members get monthly bulletins.

Immunodeficiency UK is reliant on voluntary donations. To make a donation, please go to www.immunodeficiencyuk.org/donate



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