Patients' experience survey of Primary Immunodeficiency Disorders (PID) services

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Developed by PID UK in association with CSL Behring.

PID UK
Supporting families affected by primary immunodeficiencies
Part of Genetic Disorders UK

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Foreword

The purpose of this survey was to gain valuable insight into the experiences of people affected by primary immunodeficiency disorders (PID), whether as a patient or as a family member. Empowering patients and their families to share their experiences – both good and bad – is key to influencing service improvements and shaping immunoglobulin services.

The report highlights the breadth of good work being done across the country to support people with PID, which is borne out by the survey responses to questions on participation in treatment decision-making. There is much in the report to be positive about but there are also several challenges that we need to address, including time taken to reach the correct diagnosis, lack of joined-up services and the impact of PID on people's day-to-day lives, for example, financial pressures.

In this report, we make five recommendations based on the key areas that have emerged and we will now share these widely with patients, our NHS colleagues, policy-makers, and commissioners to help us all work together to deliver the best possible care for people affected by PID.

Dr Susan Walsh, Director of Primary Immunodeficiency UK

PID UK
Supporting families affected by primary immunodeficiencies

Acknowledgements

We would like to acknowledge Interaction, a public relations and marketing consultancy company that conducted the survey, and Wicked Minds, an independent healthcare and market access consultancy that developed the patient questionnaire, analysed the data and helped prepare the report for CSL Behring UK and PID UK. We would also like to thank PID UK's patient representative panel who helped refine the questionnaire, all the patients that took part in the survey, PID UK for patient recruitment and support, and all the immunology centres that have been involved in patient recruitment.

Background

Health services in the UK support more than one million people every day[1], making the experiences of patients and their family and friends a key component of the quality of healthcare they receive. Since the Health Act came into force in January 2010, healthcare commissioners and service providers have had a legal obligation to take the NHS Constitution into account in all their decisions and actions, including focusing on patient experience[2].

The primary immunodeficiency patient experience survey was conducted between March and May 2016 and included questions relating to diagnosis, management, homecare treatment, long-term support and financial implications of living with PID. The survey gained a reliable sample size of 303 respondents from across the UK. The average response rate to the survey over this period accounts for approximately 6 per cent of the total UK PID population (5,000).

The goal of the survey conducted by PID UK, with involvement from the immunology centres, in association with CSL Behring UK is to inform NHS service improvements locally and nationally through patient insights, and ultimately deliver a better experience for patients and their carers and family members. The findings have important implications for all users of the data, and particularly for NHS policy-makers, commissioners and providers.

PID UK is an organisation supporting individuals and families affected by a primary immunodeficiency in the UK. It was established in 2013 by a grant from money raised on Jeans for Genes Day. The charity's day-to-day operations are funded entirely by donations and grants. PID UK is the UK national member of the International Patient Organisation for Primary Immunodeficiencies [IPOPI].

PID UK's aims are to help ensure that those affected by a primary immunodeficiency have the knowledge needed to manage their condition effectively and to ensure that their health needs are understood and addressed by those involved in policy and delivery of healthcare.

PID UK is a division of the charity Genetic Disorders UK (registered charity number 1141583).

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KEY FINDINGS

Gathering patient insights is a valuable source of intelligence that can help healthcare commissioners and providers make better decisions about how to improve services. Measuring patient experience is important not only to guide service improvement, but also because people's experiences of care may be linked to clinical outcomes and resource implications.

Over the course of 2 months a total of 303 responses to the patient experience survey were received - 83% completing the survey were patients and the remaining were carers/parents. The following conclusions were drawn from the results:

1. **There was significant variation in time to diagnosis from presentation of symptoms, ranging from a few months to over several years**
   - 45% of respondents reported a confirmed diagnosis taking between 1-6 years.
   - 47 respondents reported their diagnosis took between 10-20 years and 21 stated that they had been experiencing symptoms "all of their life" before diagnosis.
2. **A delayed diagnosis leads to greater burden on NHS resources**
   - 73% of respondents said that they were seen in different specialty clinics for a range of conditions or complications before confirmed diagnosis.
   - 52% of respondents stated that before confirmed diagnosis they were seen by a healthcare professional "a lot of times" (defined as over 10 times).
3. **Patients were actively involved in their treatment decision-making however there is still room for improvement**
   - 76% of respondents felt that their lifestyle, personal preference, cultural beliefs and right to choice were respected when deciding their treatment options.
   - 87% of respondents were also encouraged to participate in decisions about their health care.
   - 65% of respondents were offered choice regarding the route of immunoglobulin administration.
4. **Treatment site was dependent on route of administration**
   - 53% of respondents were on intravenous immunoglobulin therapy (IVIg) and 88% had their therapy administered in hospital.
   - 47% of respondents were on subcutaneous immunoglobulin therapy (SCIg) and 92% received their therapy at home.
5. **Treatment duration was linked to route of administration**
   - 61% of IVIg respondents received treatment every 3 weeks and 73% of SCIg respondents received treatment weekly.
   - Respondents who have their immunoglobulin treatment in hospital report it taking anything from 2 hours (23%) to half a day (20%).
6. **There may be further opportunity to offer treatment closer to home**
   - 45% of respondents are still receiving immunoglobulin treatment in hospital.
   - 97% of respondents were satisfied that they were receiving immunoglobulin therapy at the appropriate location, however 11% would still like to receive their treatment in an alternative location.
   - Of the 27 who would still like to receive their treatment in an alternative setting 85% were on IVIg as opposed to 15% who were on SCIg.
7. **A diagnosis of PID and being on long-term medical treatment can have a significant impact on a person's quality of life**
   - 40% of respondents reported that their condition and treatment had caused them to take time off work.
   - For children living with PID, 13% of respondents claimed that it caused them to take time off school.
8. **Patients reported that their condition and treatment had an emotional impact on their wellbeing**
   - 23% of respondents felt limited on their ability to travel/enjoy holidays.
   - 18% of respondents felt limited on their ability to socialise.
   - 26% of respondents claimed they had difficulties managing household activities.
   - Only 25% of respondents have access to additional specialist services or health care professionals to ensure their quality of life is maintained e.g. psychological support, social care.
9. **Regular hospital visits to receive immunoglobulin replacement therapy impose a financial burden on patients**
   - 59% of respondents faced extra expenses in the process of receiving immunoglobulin replacement therapy.
   - The most common expense incurred by 71% of respondents was for travel costs, averaging an annual expenditure of £161.
   - The largest expense incurred was for loss of wages, averaging £7,143 per annum amongst 29% of respondents.
   - 7 patients reported an average cost of £604 per year on additional medications.

Introduction

The focus on patient experience as a key element of quality in the NHS has been reinforced over the past decade through numerous reviews and policies. It was first explicitly embedded in the English NHS in 1999 as one of six domains in the NHS Performance Assessment Framework[3], which was designed to deliver high-quality, cost-effective care that would improve people's health. The experience that a person has of their care, treatment and support is one of the three parts of high-quality care first outlined by Lord Darzi in his 2008 Review[4], alongside clinical effectiveness and safety. The 2010 White Paper, *Equity and excellence: liberating the NHS*,[5] suggested that more emphasis needs to be placed on improving people's experience of healthcare and was subsequently embedded into the NHS Mandate[6] and the NHS Outcomes Framework[7], which makes clear that the provision of a 'good experience' of care for patients is a central goal for the NHS, making up one of the five core domains.

A patient's experience starts from their very first contact with the health and care system, right through to their last, which may be years after their first treatment, and can include end-of-life care. Patients' feedback on their experience of using the NHS is recognised as a key marker of the quality of those services and a vital source of information for quality improvement[8]. In 2012, NHS National Quality Board (NQB) published the NHS Patient Experience Framework, which includes an evidence-based definition of patient experience[9] and considers how this concept should be measured (Figure 1)[10]. The NQB decided that the Picker Institute framework more closely reflects the healthcare system in the UK and chose to add some additional elements around ensuring dignity, privacy and independence of service users supporting decision making and supporting self-management.

Based on a direct referral by the Department of Health (DH), the National Institute for Health and Care Excellence (NICE) published its first ever clinical guideline on *Patient experience in adult NHS services* in 2012[11]. The aim of the guidance is to provide the NHS with clear guidance on the components of a good patient experience (Figure 2). A NICE quality standard for patient experience in adult NHS services was also developed alongside this guidance[12]. NICE quality standards are a set of specific, concise statements and associated measures. The guidance draws on multiple evidence and data sources in developing the recommendations, which are further distilled into commissioning guidance in the quality standard.

'Experience' can be understood in the following ways:

1. **What** the person experiences when they receive care or treatment- for example, whether they knew who to contact if they had a problem, whether the nurse explained the procedure to them, and whether the doctor asked them what name they would like to be called by. The 'what' of people's experiences can be thought of in two ways:
    - The interactions between the person receiving care and the person providing that care, for example how a member of staff communicates with the person (this is known as the 'relational' aspects of experience);
    - The processes that the person is involved in or which affect their experience, such as booking an appointment (this is known as the 'functional' aspects of experience).
2. **How** that made them feel - for example, whether they felt treated with dignity and respect, and whether they felt that the doctor told them about their diagnosis in a sensitive way.

Figure 1. Definition of patient experience

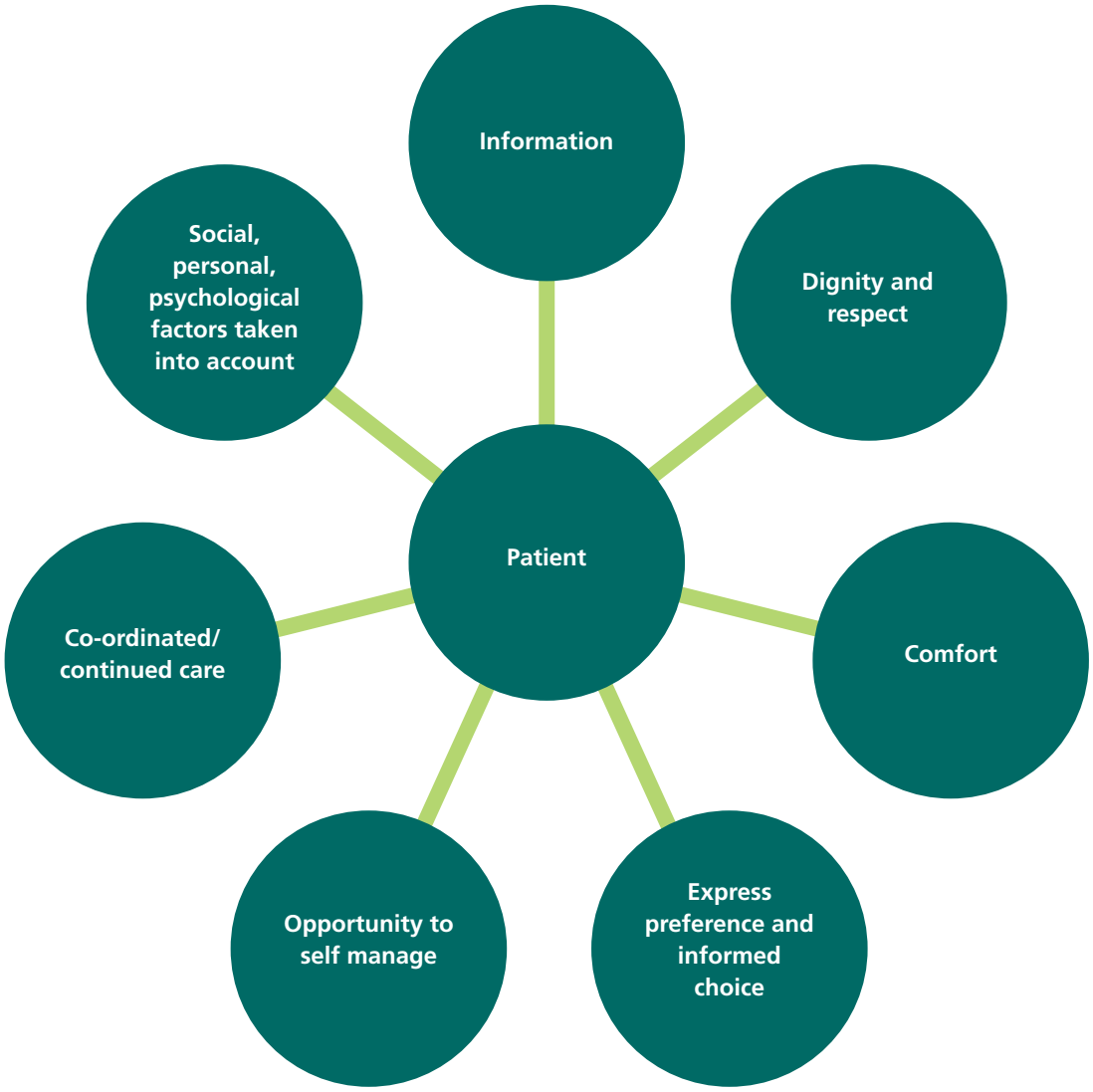


Figure 2. The outcome of good patient experience from the patient's perspective

Primary immunodeficiency disorders (PID) are a group of disorders of the immune system in which part of the immune system is missing or does not function properly. Primary denotes the mainly genetic nature of the defects, differentiating them from secondary or acquired immunodeficiencies caused by malnutrition, infection (e.g. human immunodeficiency virus [HIV] infection), chemotherapy, certain drugs or other external agents.

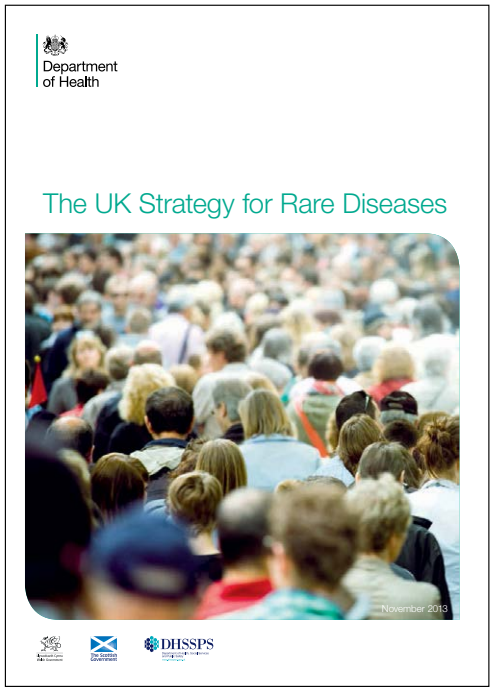
There are over 300 different but rare immune deficiencies recognised by the European Society for Immunodeficiencies[13], of which less than 20 account for > 90% of cases. These diseases range in prevalence from 1 in 3,000 to less than one in a million (overall estimate 1/50,000), and through advances in genomics the number of different types of PID identified is likely to increase. The rarity, wide spectrum, severity of complications and associated mortality and complexity of treatments require that PID be managed by immunology specialists[14].

In common with many rare diseases PIDs are frequently overlooked and reports have highlighted a diagnostic delay of up to 10 years[14] before immunoglobulin replacement therapy is started. The delay is longer in adult patients with antibody deficiency, paediatricians being more aware of the possibility of PIDs than those working in adult medicine. The delay in diagnosis contributes to increased morbidity and mortality in this patient group: overall, 25 year survival from diagnosis in this group of patients is around 75% compared to 92%

for the population of similar age, but 50% of those with complications die in a similar timeframe[14]. Early diagnosis shortens diagnostic delay that is distressing to the family, damaging to the patient and wasteful of health-care resources. About half of the patients without a diagnosis will be admitted to hospital every year; they may also be seen in different specialty clinics for a range of complications. They often receive almost continuous antibiotics for infections and are off work for long periods of time. Delayed diagnosis remains a concern for physicians and patients and continues to be an issue.

There are currently 4,168 patients registered on the UKPID Registry from 36 Immunology centres, of which 3,833 are actively attending clinics – the others are deceased, lost to follow up or discharged[15]. The national immunoglobulin demand management database reports 2,944 PID patients in England were treated with intravenous and subcutaneous immunoglobulin replacement therapy during the financial year 2014/15[16]. Immunoglobulin therapy is on the World Health Organisations (WHO) model list of essential medicines[17].

In November 2013 the DH published a UK strategy for rare diseases[18]. The strategy aimed to ensure that people living with a rare disease have the best quality of evidence based care and treatment that the health and social care systems, working with charities and other organisations, researchers and industry can provide.



The purpose of the PID UK patient experience survey is to provide information in respect of patients' diagnosis and treatment of PID, which can help the NHS to improve the quality of its health services for patients, families, and other patients living with PID.

Findings from this PID UK patient experience survey is designed to be used by multiple audiences for a range of purposes:

- By providers, to improve the quality of their services.
- By regulators, for quality assurance and to produce provider ratings.
- By commissioners, to monitor contracts and inform pay-for-performance schemes.
- By managers (locally and nationally) for performance assessment.
- By government and the public, for accountability purposes.
- By patients, so that they can make informed decisions about their care.

Data collection and methods of analysis

An online and paper based questionnaire was made available through PID UK and immunology centres to patients with PID and their caregivers regarding current treatment satisfaction, living with PID, and patient preferences (no age limit was applied). Participation was voluntary and research was conducted from March to May 2016 by an independent market research agency in accordance with ABPI, MRS and BHBIA codes of conduct towards anonymity and confidentiality. The PID patient experience questionnaire was grouped into nine 'domains' (Figure 3) of patient experience representing aspects of care that patients have said are important to them.

The questionnaire sought to establish a patient's current treatment, including route of administration, the key personnel involved in making treatment decisions, dose frequency, and site of care. It also explored patient satisfaction regarding treatment, treatment-related adverse events, and impact of PID and treatment on quality of life and financial implications. The survey took on average 30 minutes to complete and utilised rating scales, multiple choice, yes and no answers and free text.

This is the first time this type of survey has been conducted in the UK with this patient group therefore there is no baseline comparator of the patient experience over time *(please refer to appendix 1 for full survey results).*

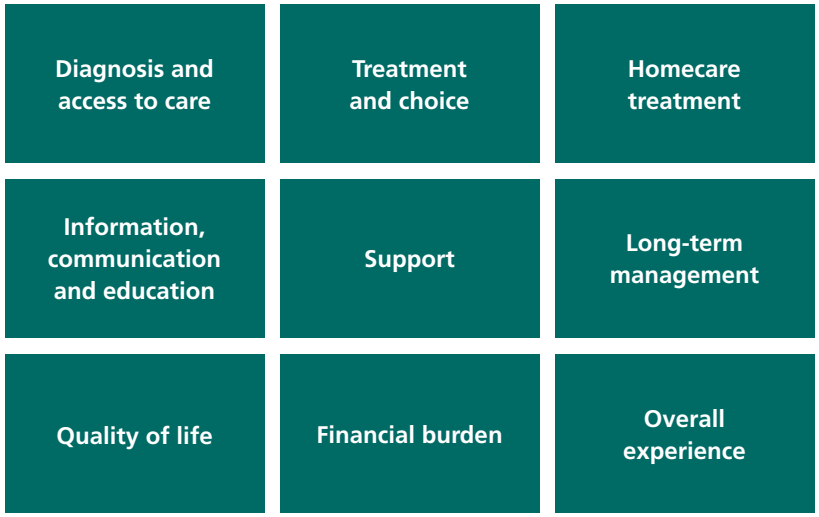


Figure 3: Nine domains of patient experience captured in this survey

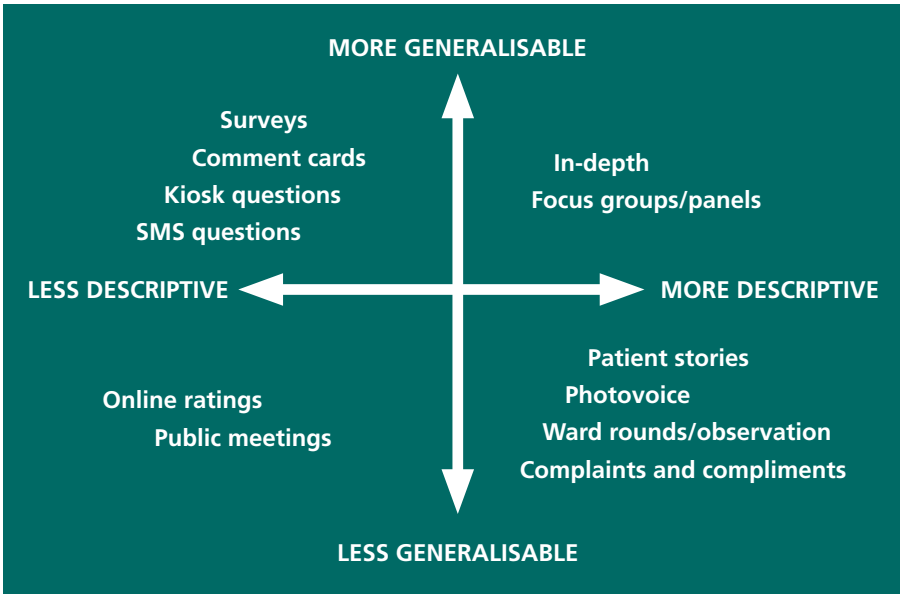


Figure 4. Examples of methods used to measure patient and carer experience of health services[2]

There are many approaches to measuring patient and carer experiences of health services (Figure 4). For the purpose of this study a survey approach was taken in order to gain a large amount of insights through multiple communication channels such as online, social media and paper based.

The limitations of a survey approach are that it only collects a surface level picture (less descriptive but more generalisable), rather than a detailed understanding of why respondents felt a certain way. In addition, patient reported experience measures (PREMS) are used to understand patients' views on their experience while receiving care, rather than the outcome of that care. Therefore, using information on both patient experience and outcomes enables a broader understanding of service quality from patients' viewpoint.

Throughout this report we have supplemented the quantitative analysis with some small-scale qualitative information that was also provided within the free text of the survey.

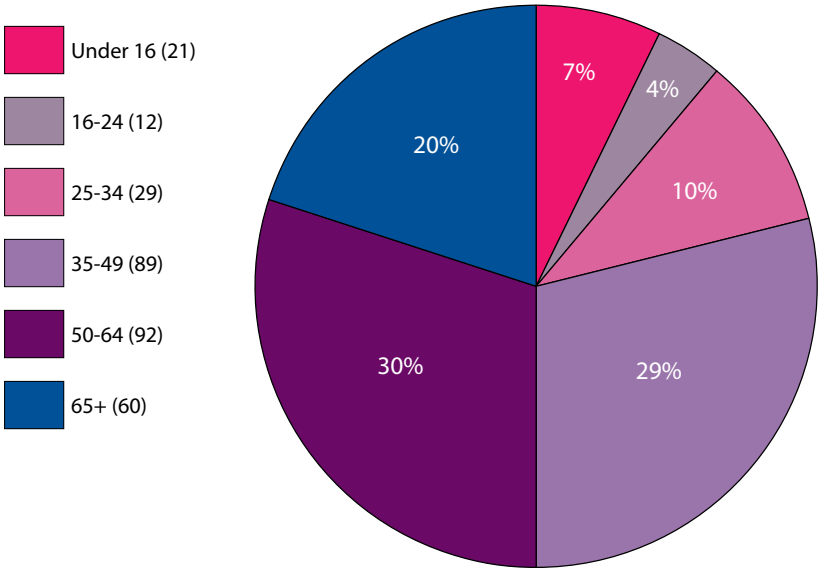
Results - Detailed analysis of survey responses

It is important to develop an understanding of the patient as an individual, including how the condition affects the person, and how the person's circumstances and experiences affect their condition and treatment. A total of 303 responded to the UK survey, 83% completing the survey were patients (250), 15% parent/carer of child (46) and 2% carer of parent/older family member (5) *(appendix 1 chart a).*

Patient demographics

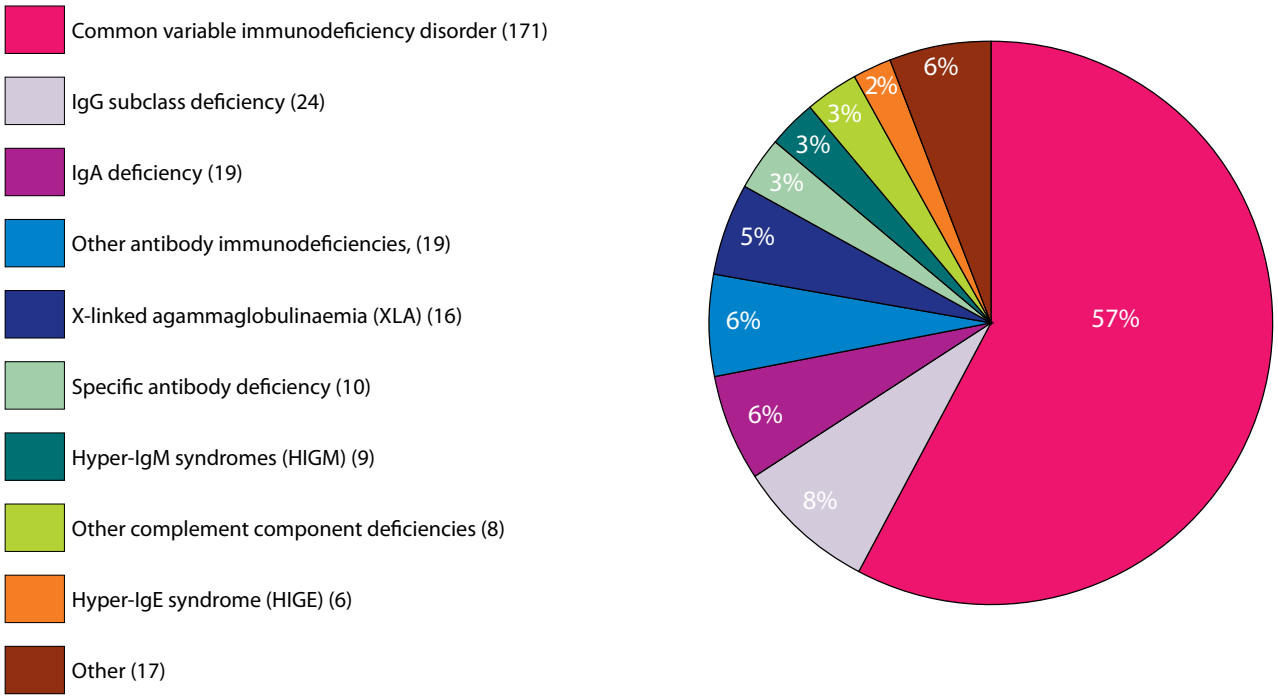
Of the 303 respondents who completed the survey, 56% (171) identified their nationality as English *(appendix 1: chart b)* and 62% (187) were female.

CHART 1: AGE OF RESPONDENTS (N=303)



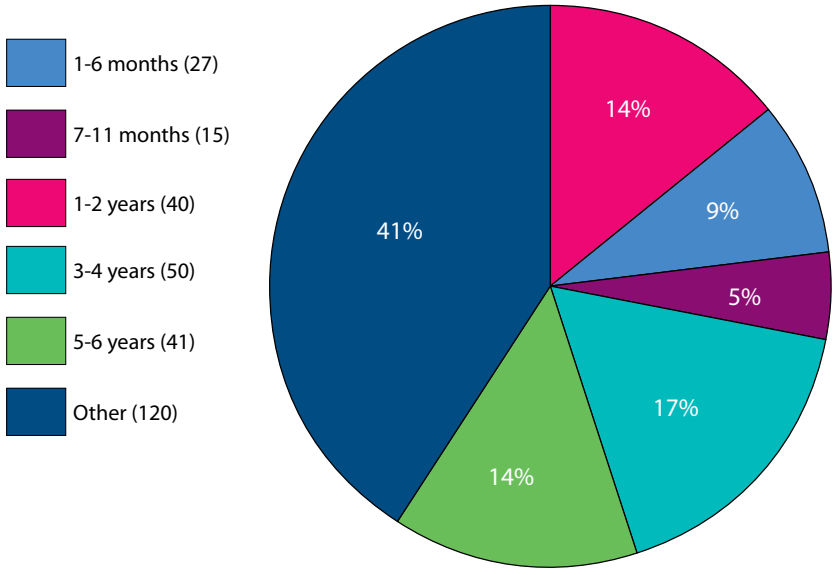
Just over half of all the respondents that completed the survey were aged between 35-64 (35-49 [29%] and (50-64 [30%]) *(chart 1)*. 57% (171) of 299 respondents classified their PID condition as a common variable immunodeficiency disorder *(chart 2)*, which reflects the majority of patients registered on the UKPID Registry.

CHART 2: HOW RESPONDENTS DEFINED THEIR IMMUNOLOGICAL CONDITION (N=299)



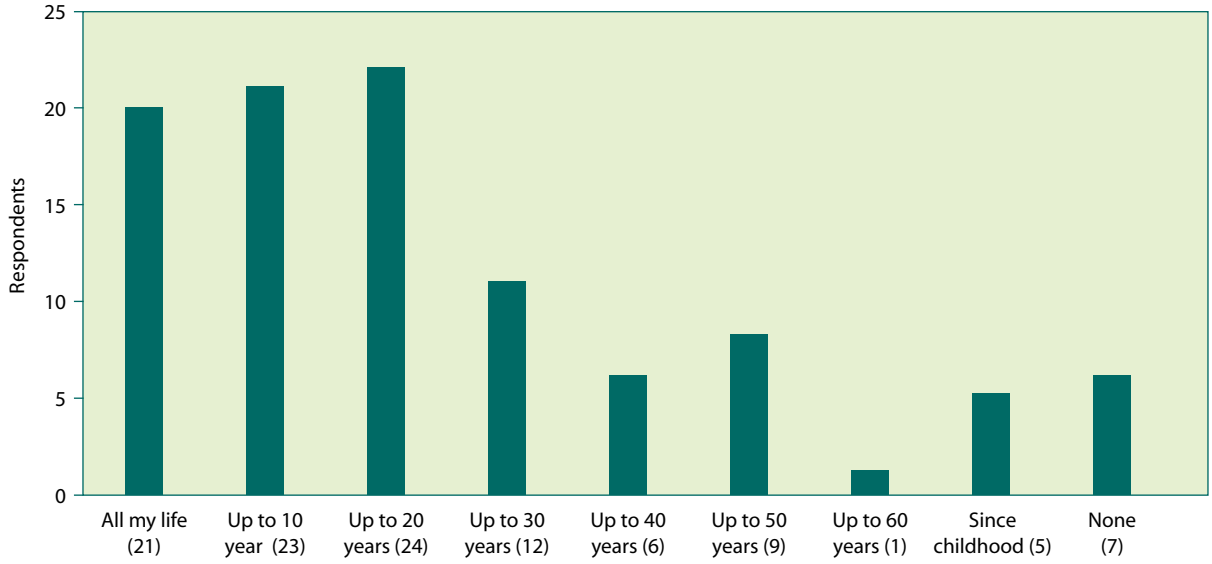
1. DIAGNOSIS AND ACCESS TO CARE

CHART 3: LENGTH OF TIME RESPONDENTS WERE EXPERIENCING SYMPTOMS BEFORE BEING DIAGNOSED (N=293)



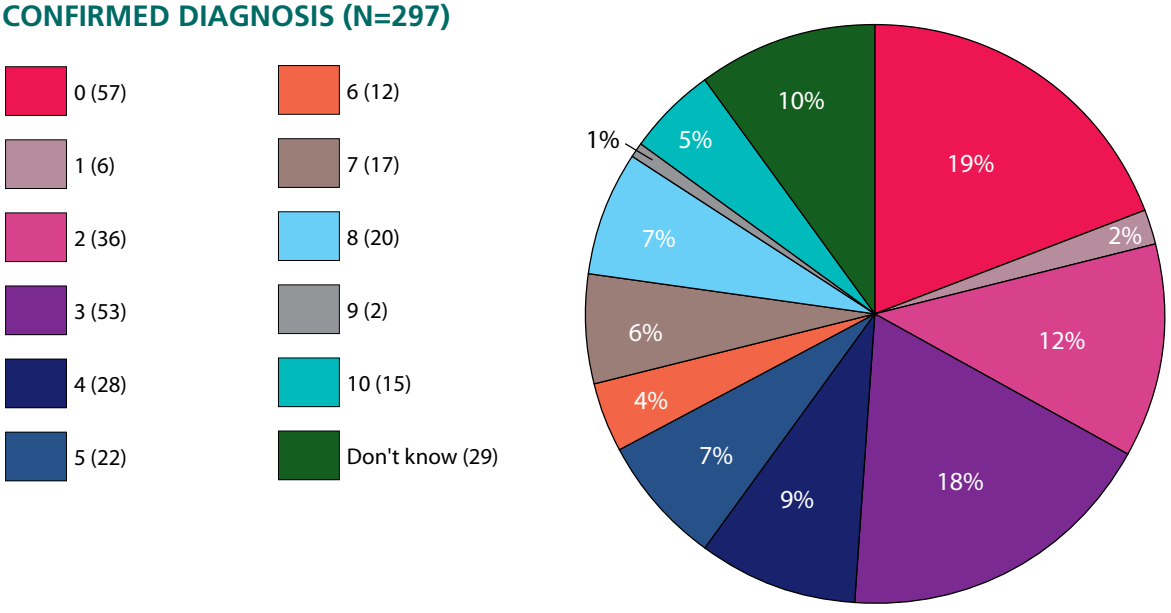
This question demonstrated a high degree of variation in time to diagnosis from presentation of symptoms ranging from a few months to over several years *(chart 3)*. 45% (131) of respondents reported a confirmed diagnosis taking between 1-6 years. 108 respondents out of the 120 (41%) who chose 'other' went on to offer additional information regarding how long they had been experiencing symptoms before being diagnosed *(chart 4)*. 47 reported between 10-20 years and 21 stated that they had been experiencing symptoms 'all of their life' before diagnosis.

CHART 4: LENGTH OF TIME RESPONDENTS WERE EXPERIENCING SYMPTOMS BEFORE BEING DIAGNOSED. "OTHER" CATEGORY EXPLAINED



The results demonstrate a significant number of years a patient remains symptomatic before a confirmed diagnosis

CHART 5: HOW RESPONDENTS FELT ABOUT THEIR SYMPTOMS BEFORE OBTAINING A CONFIRMED DIAGNOSIS (N=297)



Scale: 1 = Extremely unwell and 10 = Felt really well (average response 3.65)

68% (202) of respondents had a score of 5 or less and the average response was 3.65, suggesting that the majority of respondents felt unwell due to their symptoms prior to a confirmed diagnosis *(chart 5)*.

Many respondents referred to feeling frustrated, aggravated and annoyed regarding the amount of time it took before they were diagnosed. This has psychological implications for patients experiencing long periods of being unwell, guilty feelings by parents and increased anxiety. The direct NHS economic costs are GP time, multiple investigations, consultants' time, consultations and long period in hospital.

> *"It has been disastrous. I do not have a career and I also have been unable to start a family as I am bedridden 80% of the time. School was awful"*
>
> *Patient*

> *"The GP response was slow, however, the immunologist response was rapid with treatments starting immediately"*
>
> *Patient*

Educating GPs and primary care healthcare professionals is very important in order to know where and when to refer. If patients are not diagnosed right away the resulting direct cost to the NHS and indirect costs to the patients can be considerable. *(See appendix 1: table c)*

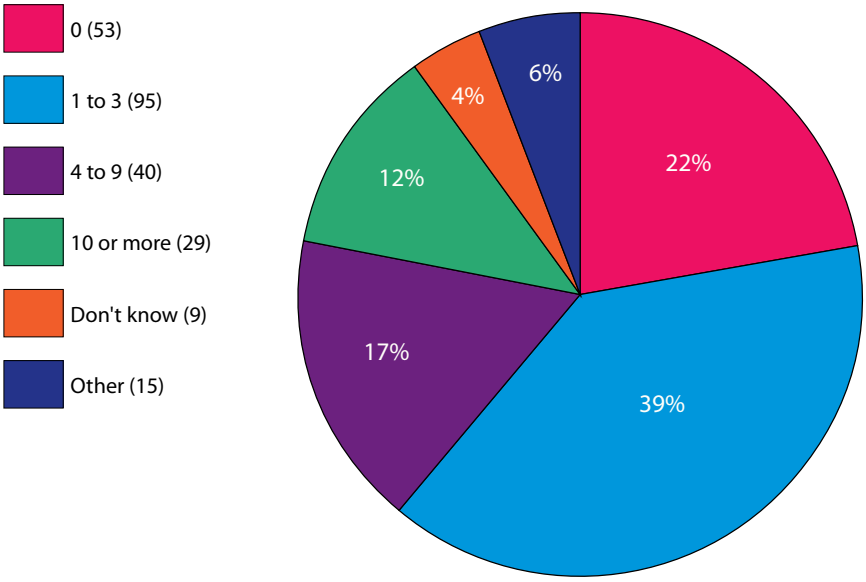
> *"It was a fluke that I even got diagnosed. I am very unhappy that a random test by a Phlebotomist was the only reason I was diagnosed. Before my confirmed diagnosis my GPs had ignored my frequent infections and medical history, treating me like I am exaggerating when I sought help from them"*
>
> *Patient*

The results showed that 60% (292) of all respondents were admitted into hospital with a condition related to their PID before they had a confirmed diagnosis. This has implications on diagnosis time and the use of hospital resources due to admissions. An average non-elective inpatient (excluding excess bed days) is £1,565[19] therefore **the cost to the NHS is £456,980** assuming all 292 respondents were only admitted once; however, 95 patients (38%) said that they were admitted to hospital between 1-3 times *(chart 6)*.

> *"It took several doctors/ consultants appointments before we had a diagnosis. It took a year of hospital stays and doctors' appointments before doctors began speaking about immunodeficiencies. I feel this could have been tested for sooner"*
>
> *Patient*

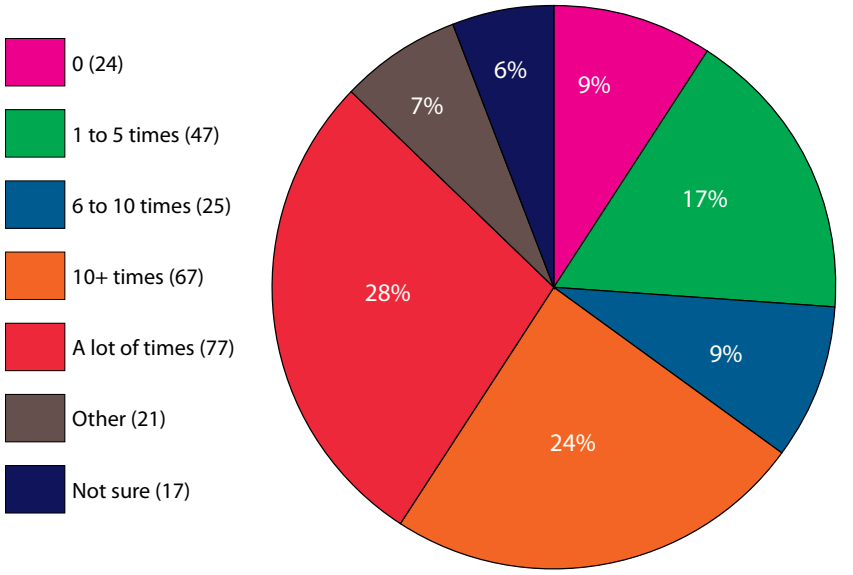
> *"Living for me ended at 60 years of age - work, marriage, everything. Deeply saddened"*
>
> *Patient*

CHART 6 NUMBER OF TIMES RESPONDENTS WERE ADMITTED TO HOSPITAL PRIOR TO OBTAINING A CONFIRMED DIAGNOSIS (N=241)



73% of respondents (212) also stated that they were seen in different specialty clinics for a range of conditions or complications before confirmed diagnosis.

CHART 7: NUMBER OF TIMES RESPONDENTS WERE SEEN BY HEALTHCARE PROFESSIONALS PRIOR TO OBTAINING A CONFIRMED DIAGNOSIS (N=278)



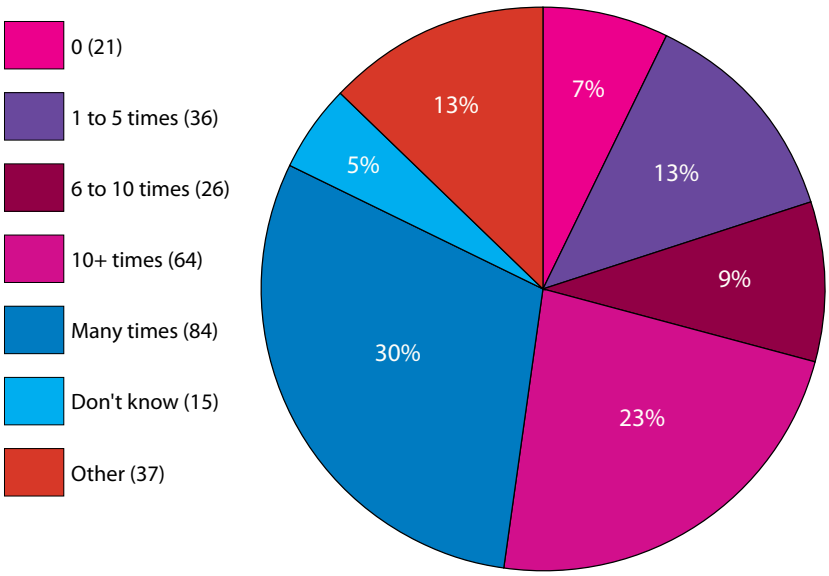
This question was asked as free text and produced a varied response from 278 respondents with 52% (24% 10+ times and 28% a lot of times) stating that before confirmed diagnosis they were seen by a healthcare professional over 10 times *(chart 7)*.

67 respondents saw a healthcare professional 10+ times before confirmed diagnosis. If these were all NHS outpatient appointments (based on an average of 10 appointments @£108[20]) it would cost the NHS £72,360 (£1,080 per patient) before a confirmed diagnosis is made.

A typical response to this question can be seen in the patient profile below:

Female patient aged 25-34 experienced symptoms for 10 years before being diagnosed. She was admitted into hospital twice and seen over 100 times by healthcare professionals in a range of specialty clinics before diagnosis.

CHART 8: NUMBER OF TIMES RESPONDENTS WERE SEEN BY A GP IN RELATION TO THEIR CONDITION PRIOR TO OBTAINING A CONFIRMED DIAGNOSIS (N=283)



This question was asked as free text and demonstrated that 30% (84) of respondents recalling 'many times' regarding how many specific times they were seen by a GP pre diagnosis. *(chart 8)*.

23% (64) respondents recall seeing the GP 10+ times which if based on 10 GP consultations lasting 11.7 minutes per surgery consultation (@£34 [excluding direct care staff costs without qualification costs] - @£44 [including direct care staff costs with qualification costs][21]) would equate to 64 patients costing the NHS between £21,760 - £28,160.

Before the diagnosis is made, an individual suffering from recurrent bouts of infections, autoimmune, or inflammatory disease due to PIDs is often investigated by many different specialists but without appropriate treatment or management. The end result is deterioration of the patient's condition, inappropriate use of health resources, and a feeling of helplessness among all parties. In general, the earlier the underlying diagnosis of a PID is made, the less damage (e.g. lung complications) will be done and, often, the easier it is to treat and manage the disease successfully. Doctors and nurses are often not familiar with the characteristic signs and symptoms of a PID and this can, in some cases, lead to significant diagnostic delay. An accurate diagnosis of a PID may also be delayed because the primary symptom of the disorder, a series of infections, can easily be attributed to 'ordinary' illness, especially in children. Frequent infections are common among children, even those who would otherwise be classified as 'healthy'. For an adult patient showing signs of a PID, there are other factors in diagnosis delay. While severe cases of a PID are typically diagnosed in children, symptoms revealing milder forms often do not appear until later in life. In addition, **sometimes health professionals just don't pick up on the pattern and frequency of infections, which is necessary to quickly make the proper diagnosis**. An example of this is upper and lower respiratory tract infections which are often attributed to something else[22]. Pilot studies are underway to provide possible solutions to reducing diagnostic delay, for example; the potential to use low levels of calculated globulin to detect antibody deficiency.

The NHS Patient Experience Survey Framework highlights 'Information, communication, and education' as an important element of patient experience, outlining that patients should be informed on clinical status, progress, prognosis, and processes of care in order to facilitate autonomy, self-care and health promotion. In this survey, 73% (205) of respondents received a copy of the letter sent to GP on diagnosis, which demonstrates that the specialist centre had kept the GP informed on the patient's diagnosis enabling greater continuity of care across the care providers. Likewise, 27% of patients did not receive a copy of the letter sent to the GP and it is unclear whether this was just a process issue or whether the GP had been informed at all. **It is essential that all centres work to an agreed standard** (NHS England CRG service specification and Royal College of Physicians [RCP] Quality Performance Indicator Data [QPID] criteria for gaining accreditation) to ensure there is effective communication and integration of care, and patients' feel empowered and informed to be actively involved in their care plan.

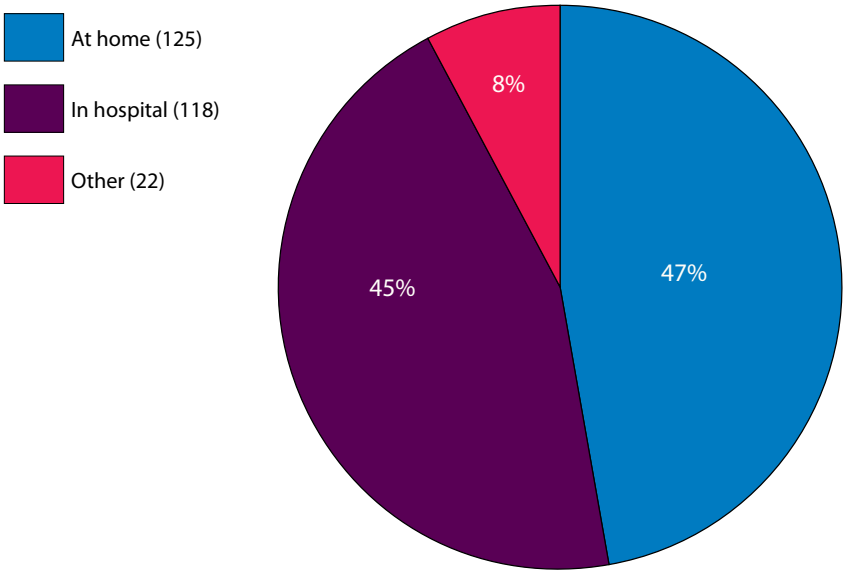
2. TREATMENT AND CHOICE

Patient choice is at the heart of the NHS. There are a number of choices that patients should expect to be offered across the range of NHS services on offer. The Choice Framework, published by the DH, sets out the nine main types of choices that should be available to patients in the NHS[23]. Some of these choices are legal rights, while some are subject to specific exceptions. Survey evidence[24] shows that progress towards achieving meaningful choice for NHS patients has stalled over recent years. NHS England is therefore committed to a major programme of work to realise the NHS' longstanding promise to give patients choice over where and how they receive care, as highlighted in the NHS Five Year Forward View[25].

In line with the NICE guideline [CG138][11] on *Patient experience in adult NHS services: improving the experience of care for people using adult NHS services*, it is encouraging to see that 76% (230) of respondents felt that their lifestyle, personal preference, cultural beliefs and right to choice were respected when deciding their treatment options *(appendix 1: chart d)*. 87% (232) of respondents were also encouraged to participate in decisions about their health care.

53% (134) of respondents who completed the survey were on IVIg therapy and of these 15 were receiving their treatment at home as opposed to 107 in a hospital setting (2 did not answer and 10 responses were noted as 'other'). 47% (121) of respondents were on SCIg replacement therapy majority (109) of whom were receiving their treatment at home (10 respondents received SCIg in hospital and 2 responses were noted as other). *(appendix 1: chart e)*.

CHART 9: WHERE RESPONDENTS STATED THAT THEY CURRENTLY RECEIVE THEIR IMMUNOGLOBULIN TREATMENT (N=265)



The Five Year Forward View[25] has encouraged efforts to deliver more healthcare out of acute hospitals and closer to home, with the aim of providing better care for patients, cutting the number of unplanned bed days in hospitals and reducing net costs. Of the 265 respondents 45% are still receiving immunoglobulin treatment in hospital. The NHS agenda is about driving treatment closer to patient's home, however there were 118 respondents coming into hospital for long-term immunoglobulin treatment *(chart 9)*.

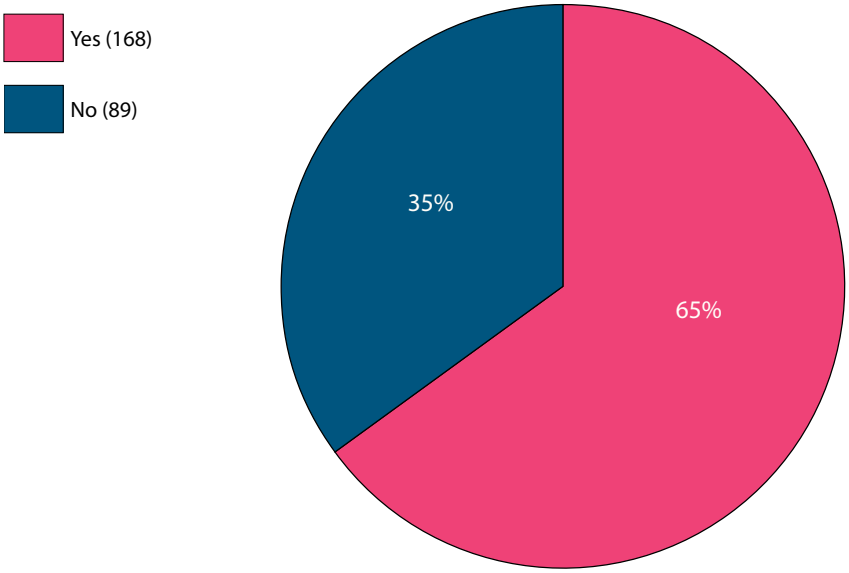
Out of the 22 respondents that stated 'other':

- 6 had not started on immunoglobulin replacement therapy yet.
- 1 respondent has their treatment at a friends.
- 1 respondent has their treatment at the GP.
- 1 respondent has their treatment at the day clinic.
- 10 respondents do not have immunoglobulin replacement therapy.
- 3 respondents did not state a reason.

The advent of home therapy programmes has given many patients a better quality of life as it is more convenient than receiving infusions at hospital and allows patients to have a greater feeling of control over their condition. All centres should offer a home treatment service for long-term immunoglobulin replacement consistent with the recommendations of the National Immunoglobulin Demand Management Programme, with appropriate monitoring, training and governance. Potential benefits for all patients include cost–efficacy: avoids hospital episodes (outpatient treatment and hospital admission and promotes home care), better care to current standards: major improvement in quality of life and reduced variation in care and access.

Lord Carter's review on *'Operational Productivity and Performance in English NHS acute hospitals'*[26] recommended that Trusts should ensure more clinical pharmacy staff are deployed – working more closely with patients, doctors, nursing staff and independently – to deliver optimal use of medicines, make informed medicines choices, secure better value, drive better patient outcomes, and contribute to delivering 7 day health and care services. It went on to highlight that pharmacy staff should focus further in improving productivity and efficiency, including consideration of alternative supply routes, such as homecare providers or community pharmacies.

CHART 10: RESPONSES TO WHETHER RESPONDENTS WERE OFFERED A CHOICE BETWEEN INTRAVENOUS AND SUBCUTANEOUS THERAPY (N=257)



The NHS constitution[27] says that patients have the right to choose treatment. SCIg and IVIg replacement therapy is the mainstay of treatment for most patients with PID. Patient preference studies show that subcutaneous therapy is preferred by patients and has the potential to help alleviate nurse shortages and reduce overall health care costs[28]. The survey showed that 35% (89) of the 257 respondents failed to be offered choice of mode of administration, however the reasons for this were not explored *(chart 10)*.

Of the 134 respondents on an IVIg, 63% (85) were offered a choice, 34% (46) were not and 2% (3) did not comment. Of the 121 respondents on SCIg, 67% (81) were offered a choice, 31% (38) were not, and 2% (2) did not answer. In addition, out of the total respondents 23% (60) were not offered a choice of where they would like to receive their treatment (i.e. home or hospital) *(appendix 1: chart f)*. Of those 60 who were not offered a choice, 53% (32) were on IVIg and 42% (25) on SCIg – 3% (3) did not specify what treatment they were on. **This goes against the requirements of the national immunology service specification[14] which states that the provider shall provide patients the option of home therapy.**

Studies such as Kittner *et al* (2006)[29] have demonstrated that patients on SCIg therapy were significantly younger than those in the IVIg treatment group (37±9.1 years versus 51.2±14.5 years, P<0.001). Another study investigated the perceptions of the subcutaneous method and the perceptions of the home therapy regime among patients who already had considerable experience of this therapy - significant correlations were found between age and perception of the lifelong treatment with SCIg infusion of immunoglobulin for primary antibody deficiencies[30]. This patient experience survey did not demonstrate any age related variation to treatment choice, however the patient pool from this survey is not representative of all age groups as just over half of all the respondents that completed the survey were aged between 35-49 (29%) and 50-64 (30%).

CHART 11: RESPONDENTS AGE IN RELATION TO TREATMENT CHOICE OF EITHER INTRAVENOUS (N=134) OR SUBCUTANEOUS THERAPY (N=121)

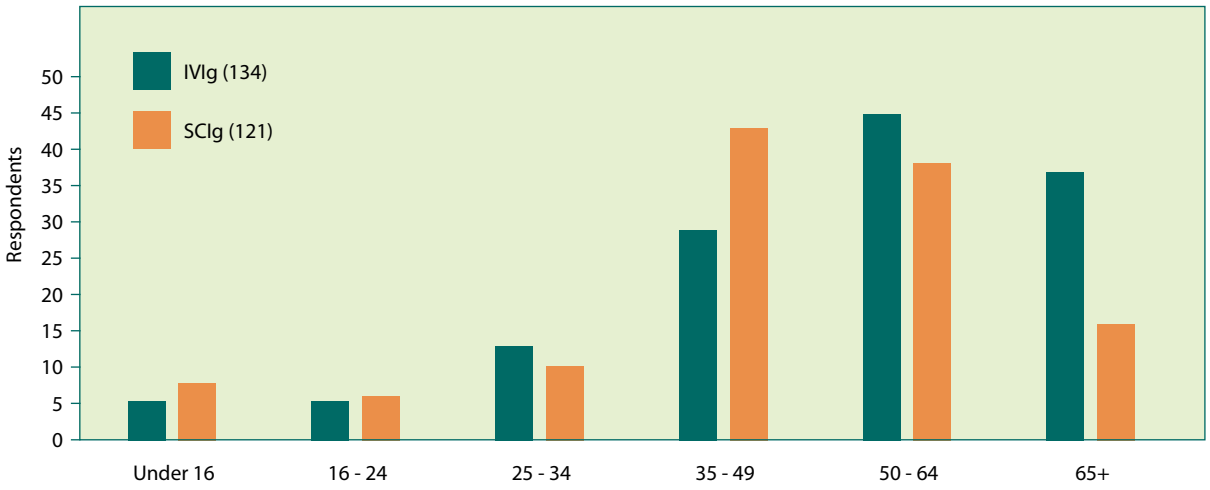
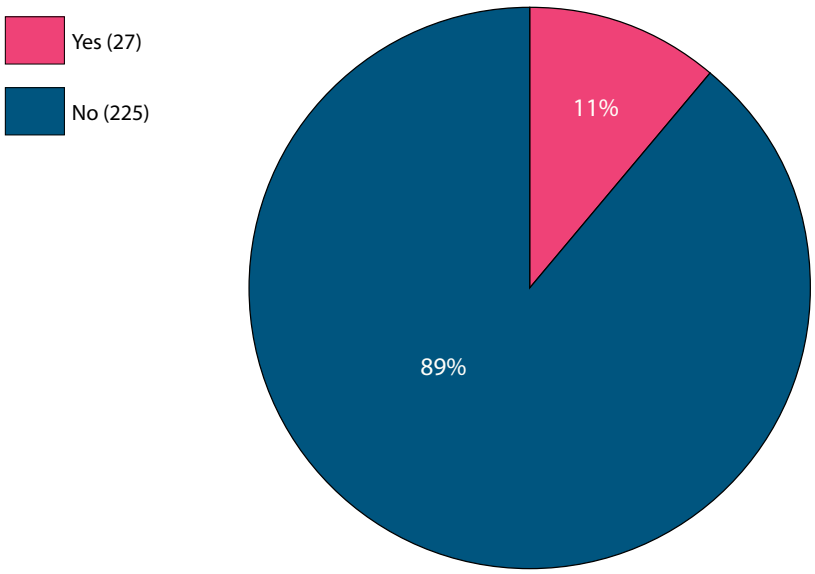


CHART 12: RESPONDENTS VIEWS ON WHETHER THERE IS ANY OTHER LOCATION THAT THEY WOULD PREFER TO RECEIVE TREATMENT (N=252)



97% (247) of respondents were satisfied that they were receiving their immunoglobulin therapy at the appropriate location *(appendix 1, chart g)*, however 11% (27) would still like to receive their treatment in an alternative location *(chart 12)*. Interestingly, of the 27 who would still like to receive their treatment in an alternative setting 85% (23) were on IVIg as opposed to 15% (4) who were on SCIg.

> *"It would be good if I could be cannulated at my GP surgery and not have to travel into hospital. However, I don't think there are staff with the right skills and there are issues of time and funding etc"*
>
> *Patient*

> *"Currently we travel to St. Georges which is about a 35 minute drive from Epsom. We would love to have the transfusion at Epsom General which is our local hospital and about a 7 minute drive away"*
>
> *Family member/Carer*

> *"Home, if my GP ever agrees"*
>
> *Patient*

"You have the right to receive care and treatment that is appropriate to you, meets your needs and reflects your preferences." (Section 3a of the NHS Constitution[26]). These survey results suggest that more needs to be done to improve patient choice of treatment and the location in which the patient prefers to receive their treatment.

CHART 13: NUMBER OF TIMES RESPONDENTS WERE GIVEN THEIR IMMUNOGLOBULIN TREATMENT IRRESPECTIVE OF WHETHER THEY ADMINISTERED IT THEMSELVES OR VIA A HEALTHCARE PROFESSIONAL (N=262)

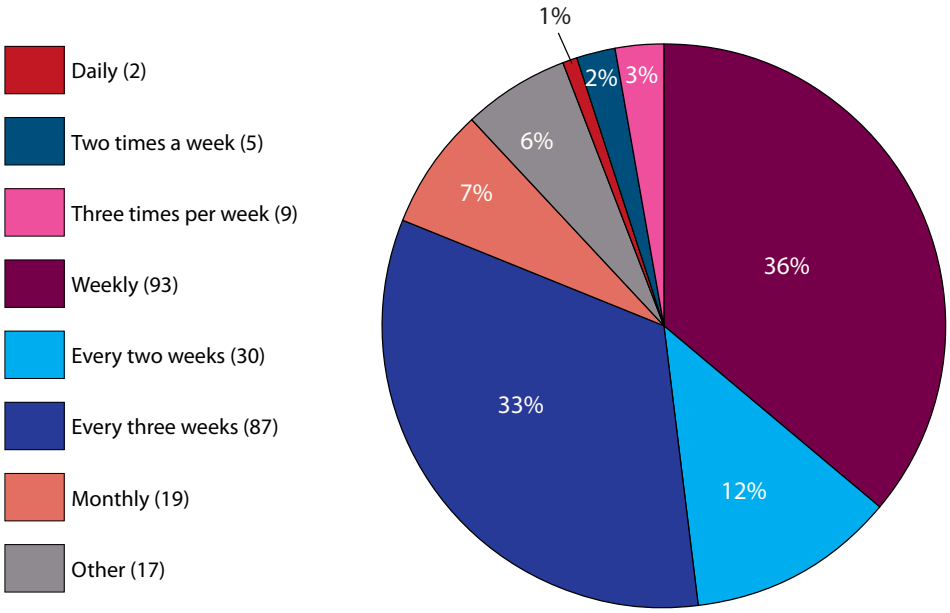
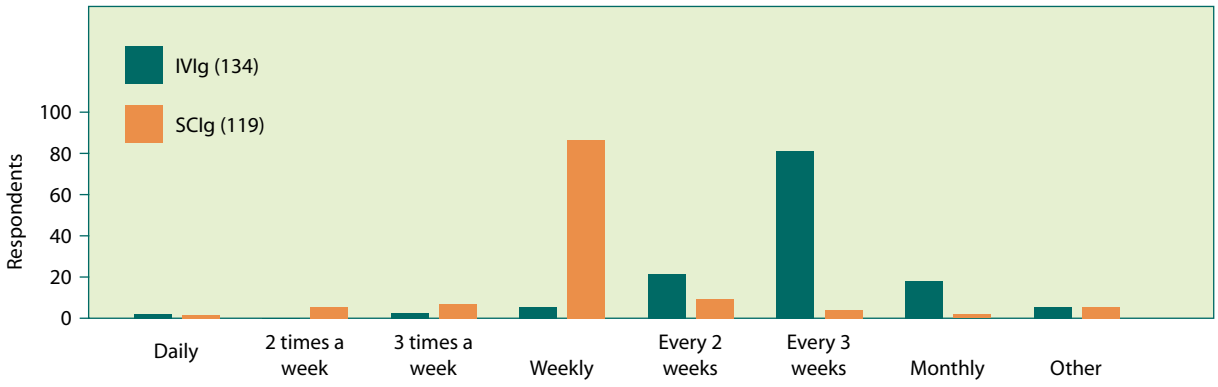


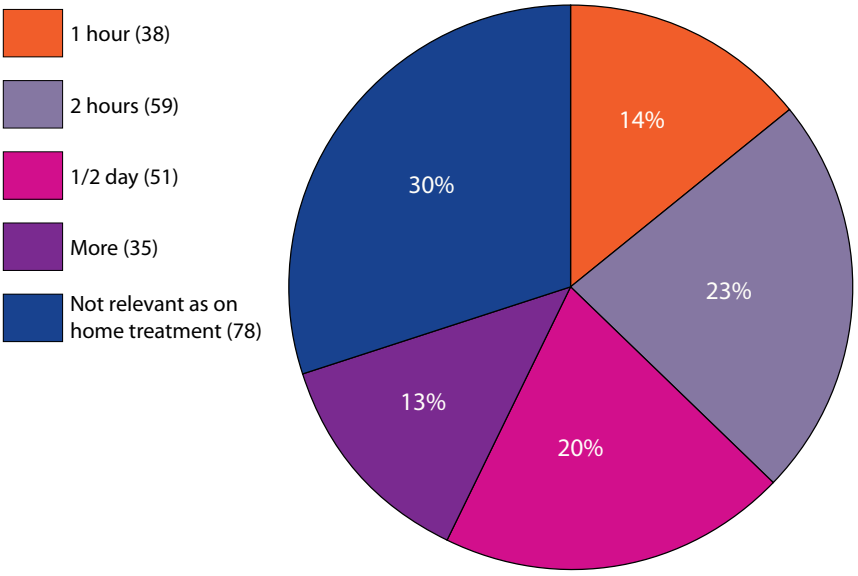
CHART 14: NUMBER OF TIMES RESPONDENTS WERE GIVEN THEIR IMMUNOGLOBULIN TREATMENT SPLIT VIA IVIG (N=134) AND SCIG (N=119)



36% (93) of respondents have their immunoglobulin treatment weekly whilst 33% (87) of respondents have their immunoglobulin treatment every three weeks *(chart 13)*. The main distinction between the SCIg and IVIg treatments is the frequency of infusions (chart 14). Typically, IVIg patients receive infusions every three weeks, whereas SCIg patients infuse every week. In fact, some SCIg patients are opting to infuse even more frequently (2-3 times a week). For instance, some doctors recommend patients give themselves one small daily injection instead of a single larger weekly infusion. Daily infusions are given by push, rather than with a pump. The advantage of giving immunoglobulin treatment by push several times a week is the simplicity of the procedures and the little time it takes.

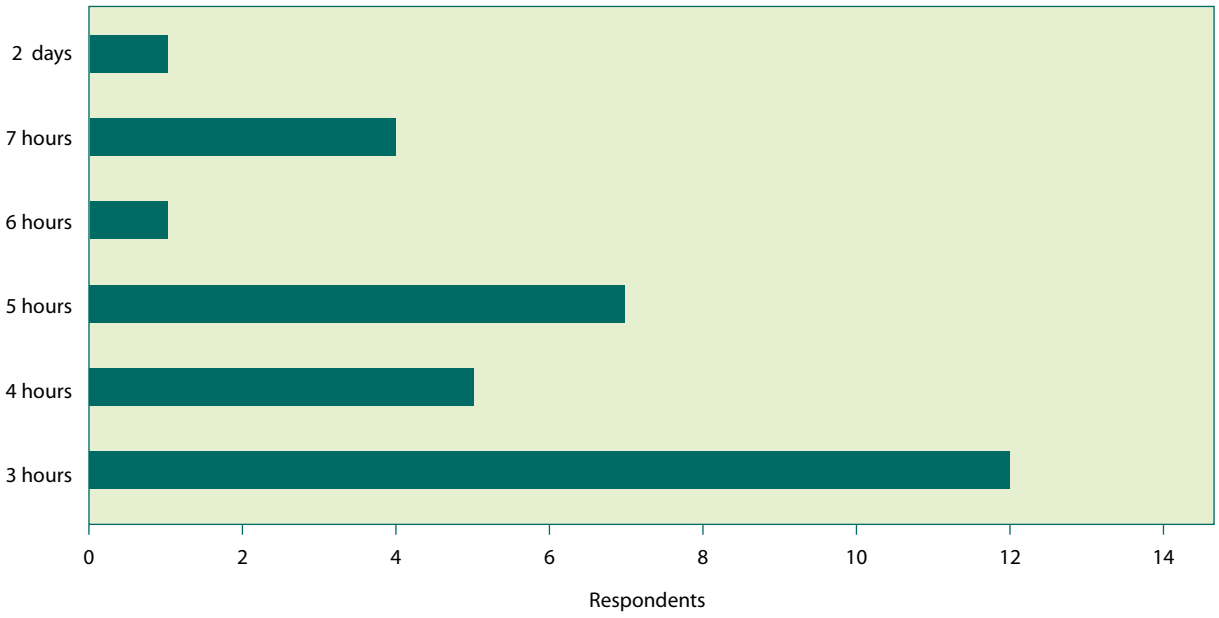
37% of respondents report to having a healthcare professional taking blood samples once every 3 months as part of managing their treatment regime *(appendix 1: chart h)*.

CHART 15: LENGTH OF TIME EACH TREATMENT REGIME TAKES. INCLUDES TRAVEL TO AND FROM HOSPITAL, SET UP AND ADMINISTRATION (N=261)



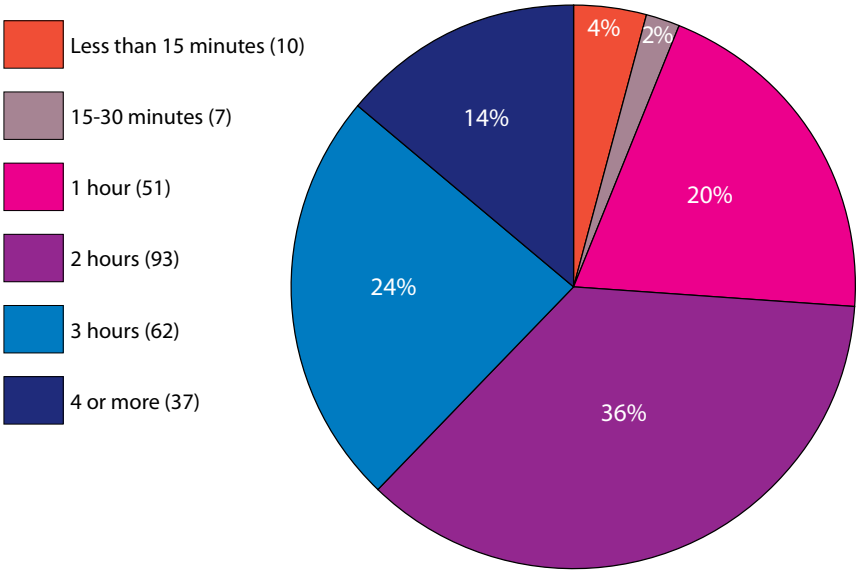
Respondents who have their immunoglobulin treatment in hospital report it taking anything from 2 hours (23% [59]) to half a day (20% [51]) *(chart 15)*. Of those who said 'more' (35) 30 respondents gave answers ranging from 3 hours to 2 days *(chart 16)*.

CHART 16: "MORE" CATEGORY EXPLAINED (N=30)



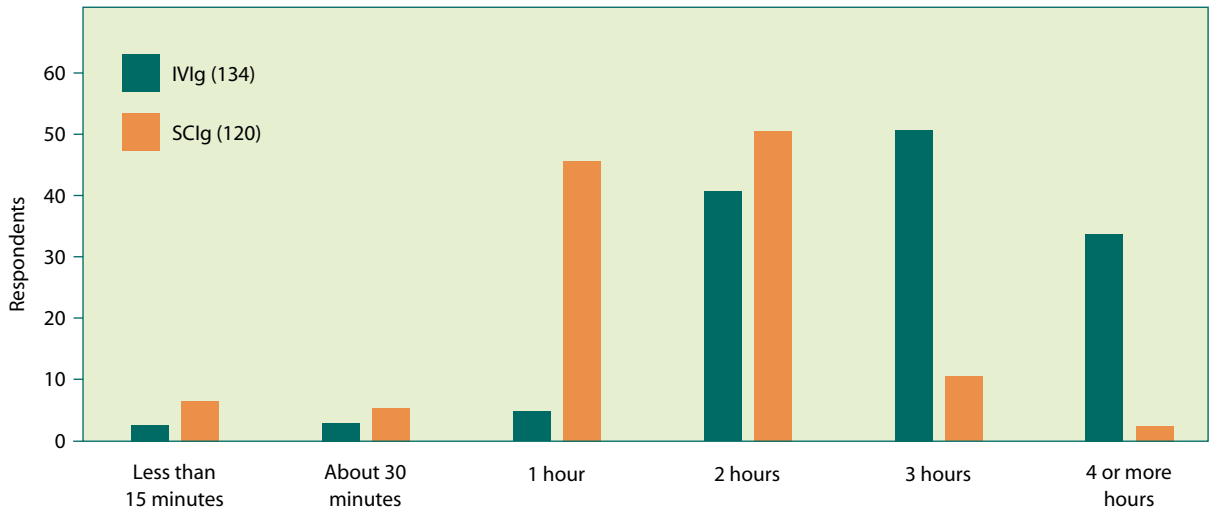
Given the long travel and hospital time for each treatment regime it is important to understand that 45% (113) of respondents do not have to take annual leave to attend appointments, however there are still 20% of respondents (51) who do *(appendix 1: chart i)*.

CHART 17: DURATION OF EACH TREATMENT REGIME (N=260)



Duration of each treatment regime is varied and there is wide variation ranging from up to 30 minutes in 6% (17) of respondents, 2 hours for 36% (93) and 4 or more hours for 14% (37) of respondents *(chart 17)*.

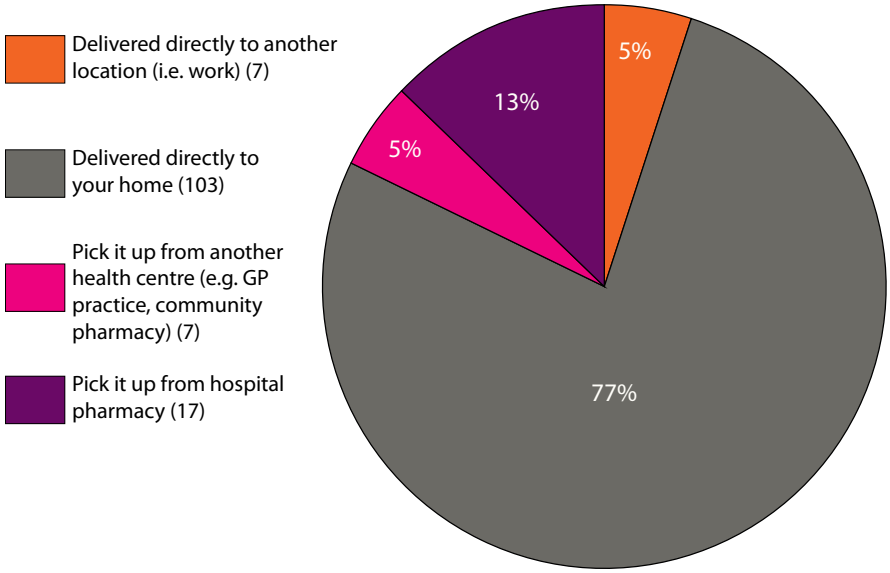
CHART 18: DURATION OF EACH TREATMENT REGIME SPLIT BY MODE OF ADMINISTRATION



3. HOMECARE TREATMENT

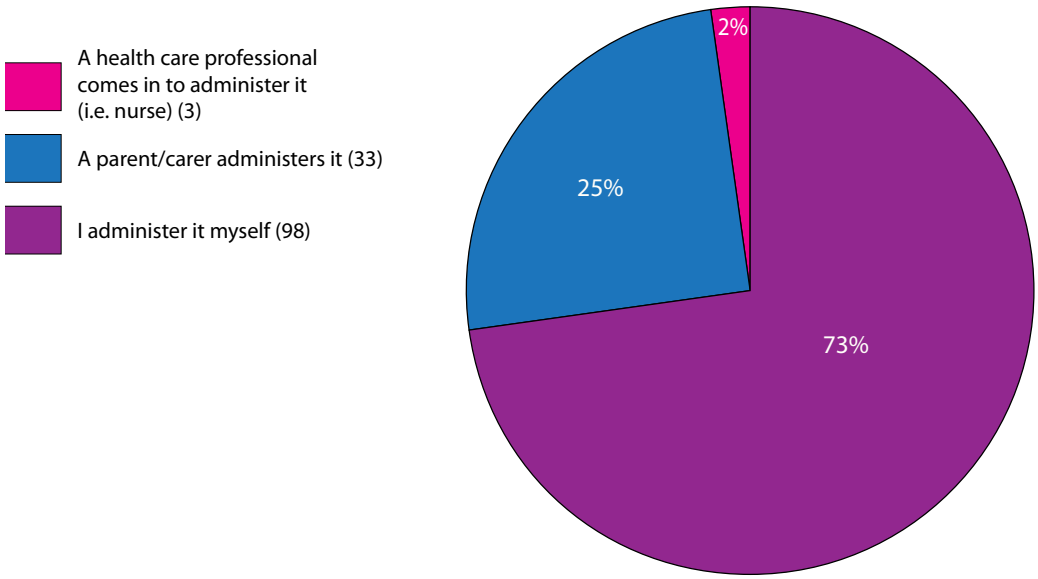
Home therapy programmes for patients requiring IVIg or SCIg are already available in the majority of immunology centres but not comprehensively delivered. It is recognised that managing chronic conditions outside of the hospital environment improves patients' quality of life. At the same time, it allows hospital services to adapt to meet new service demands without creating a burden for community health services.

CHART 19: HOW RESPONDENTS' IMMUNOGLOBULIN TREATMENT IS ACCESSED FROM HOME (N=134)



The majority of respondents (77% [103]) have their immunoglobulin delivered directly to their home, however 13% (17) of respondents who are on homecare treatment are still travelling to their hospital to pick it up from pharmacy *(chart 19)*.

CHART 20: WHO ADMINISTERS THE RESPONDENTS IMMUNOGLOBULIN TREATMENT AT HOME (N=134)



32% (98) of respondents administer the immunoglobulin replacement therapy themselves *(chart 20)* with the majority 47% (62) attending on average 2-3 sessions to be trained to self-administer *(chart 21)*.

CHART 21: NUMBER OF TRAINING SESSIONS RESPONDENTS ATTENDED IN ORDER TO BE ABLE TO SELF-ADMINISTER IMMUNOGLOBULIN TREATMENT (N=131)

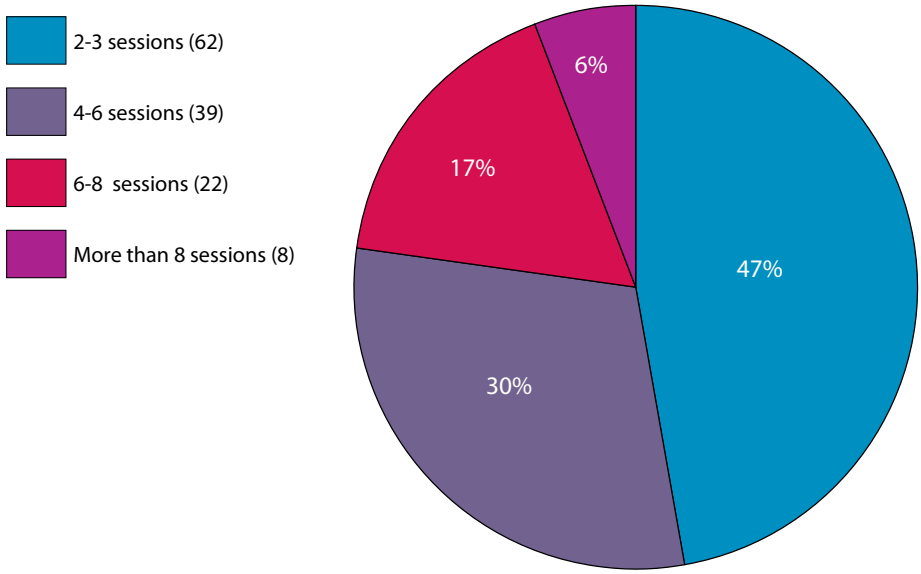


CHART 22: NUMBER OF TRAINING SESSIONS ATTENDED IN ORDER TO BE ABLE TO SELF-ADMINISTER IMMUNOGLOBULIN TREATMENT BY TYPE OF RESPONDENT (N=130)

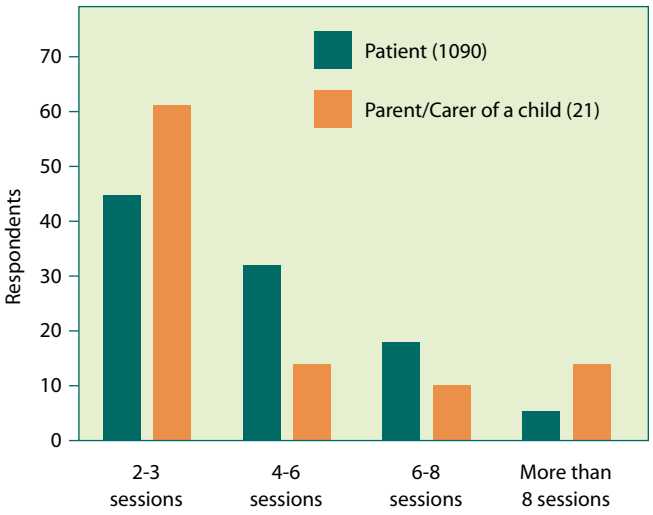
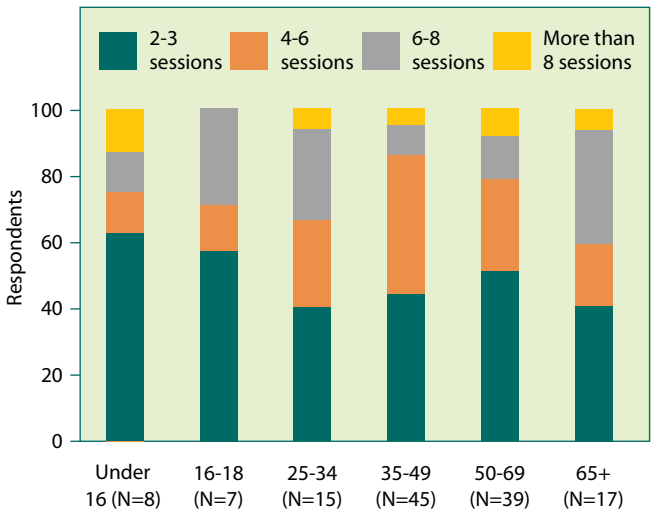


CHART 23: NUMBER OF TRAINING SESSIONS ATTENDED IN ORDER TO BE ABLE TO SELF-ADMINISTER IMMUNOGLOBULIN TREATMENT BY RESPONDENT AGE CATEGORY (N=130)



80% (104) of respondents said that their ability to self-administer had been tested after receiving the home therapy training *(appendix 1: chart j)*, however, only 43% (131) answered the question showing that 18% (24) had not been tested for competence. This may have patient safety implications and is a key part of the CRG specification and QPID accreditation scheme; and suggests some centres are not working to agreed standards.

95% (125) of respondents had been provided with the relevant infusion sets, pumps for subcutaneous delivery and deliverables to their home. 30 respondents expressed they have concerns in connection with the equipment and/or consumables delivered to their home *(appendix 1: table k)*.

> *"1 in every 3 orders has something wrong, requiring me to chase them and get correct equipment sent or return equipment I don't need"*
>
> *Patient*

> *"Sometimes the frequency is too often and I have surplus consumables. Full sharps bin is not always collected by driver"*
>
> *Patient*

> *"I have to collect all my equipment from three different locations, none of which are within walking distance and I don't drive. Obviously when you are regularly sick this can mean it is difficult to travel on public transportation which is full of germs and it is then hard to carry large loads home again"*
>
> *Patient*

> *"The delivery was quite difficult as at first only part of the equipment had been provided leading to booking a last minute hospital session. Then it was not for the correct number of months. Finally, the needles bin has never been collected after 7 months now, therefore, I feel the company who provides the medicine and the equipment could be a bit better organised"*
>
> *Patient*

In recent years there have been concerns around the governance protocols for homecare provision. In April 2014, NHS England issued an alert to NHS Trusts stating that reports of drugs failing to be delivered on time had 'increased significantly'. It instructed Trusts to put alternative methods of supply in place for patients whose deliveries were delayed and to assess providers' capabilities before assigning them more patients.

> *"I LOVE having the freedom and independence to do this from home. The training was really practical and empowering"*
>
> *Patient*

82% (108) of respondents did state that their homecare treatment is reviewed regularly *(appendix 1: chart l)*, however there is **wide variation in the definition of 'regularly'** ranging from 36% (39) having yearly reviews to 31% (33) twice a year and 23% (25) quarterly *(appendix 1: chart m)*.

4. INFORMATION, COMMUNICATION AND EDUCATION

The survey addressed questions around information, communication and education on clinical status, progress and prognosis; on processes of care; to facilitate autonomy, self-care and health promotion in line with the NHS Patient Experience Framework. 91% (248) of respondents said that they were given clear information either written or verbal and 87% (235) felt that they were given enough time to be involved in decision making about their treatment. Less positive was that just over half of the respondents 61% (166) were made aware by their healthcare professional as to the impact that living with a PID may have on everyday activities. *(appendix 1: chart n)*.

Principle 4 of the NHS Constitution[26] states that: The patient will be at the heart of everything the NHS does The NHS should support individuals to promote and manage their own health. NHS services must reflect, and should be co-ordinated around and tailored to, the needs and preferences of patients, their families and their carers.

Healthcare providers should support co-ordinated care through clear and accurate information exchange (written and shared through a personalised care plan) between patient and relevant health and social care professionals. Where appropriate patients and carers should be signposted to patient organisations such as PID UK that can provide further advice and support with their ongoing needs.

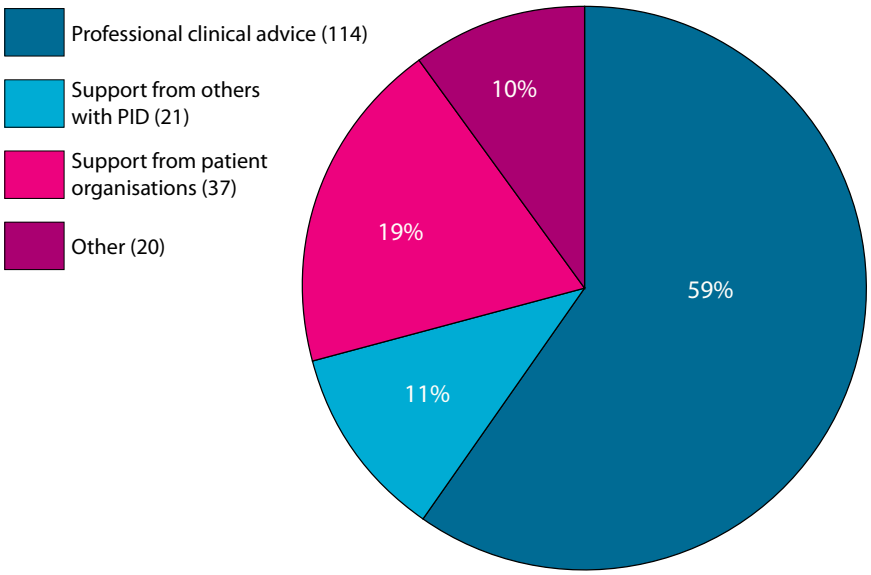
5. SUPPORT

The survey addressed questions on the different preferences for support:

- Support for self-care and individual coping strategies.
- Education.
- Need for emotional support.
- Responsiveness of health care professionals to individual support needs.
- Importance of peer-support, groups, voluntary organisations.
- Practical support.
- Family and friends support.
- Role of advocacy.

31% (85) of respondents were NOT aware of available support to help them overcome any fears or anxiety relating to their PID condition and treatment that they may experience *(appendix 1: chart o)*. Out of those who were aware (192) 59% of support was professionals' clinical advice *(chart 24)*.

CHART 24: TYPES OF SUPPORT RESPONDENTS RECEIVED IN ORDER TO HELP ALLEVIATE ANY FEARS OR ANXIETY RELATING TO THEIR PID TREATMENT (N=192)



Respondents were asked to tick from a multiple-choice list any that would help them to manage their condition better. The top 3 were: written disease-specific information leaflets (39%), more information about available therapy/treatment options (37%) and regular educational events for patients (37%) *(appendix 1: chart p)*. This response offers an opportunity to co-produce support tools with people living with PID to help future patients manage their condition better.

54% (150) of respondents stated that they are NOT in contact with other patients who have PID and of those 46% (69) would like to be in touch with other patients.

> *An opportunity to talk specifically about any benefits and employment issues*
>
> *Patient*

> *Materials for schools. My son's school doesn't take it seriously*
>
> *Family member*

> *Explanatory information for friends, family, employers to outline potential implications on relationships and ability/performance*
>
> *Patient*

> *GP and others to be made more aware of my condition in order for them to understand that I need longer courses of antibiotics when I am sick*
>
> *Patient*

> *I'm currently pregnant and would enjoy discussing living with PID and coping with pregnancy*
>
> *Patient*

6. LONG-TERM MANAGEMENT

83% (223) of respondents reported that they had discussions regarding any problems or complications to look out for following their PID diagnosis (e.g. recurrent infections, obtaining or taking antibiotics). This is important as 61% (163) stated that they had secondary complications as a result of their immune deficiency. 93% (250) were clear about what to do if any problem occurred and who to contact.

CHART 25: NUMBER OF REGULAR OUTPATIENT CLINICS ATTENDED BY RESPONDENTS IN THE PAST 12 MONTHS FOR ASSESSMENT AND FOLLOW-UP (INCLUDING MONITORING OF ANTIBODY LEVELS, BLOOD COUNTS AND LIVER FUNCTION TESTS) RELATING TO THE MANAGEMENT OF THEIR PID CONDITION (N=275)

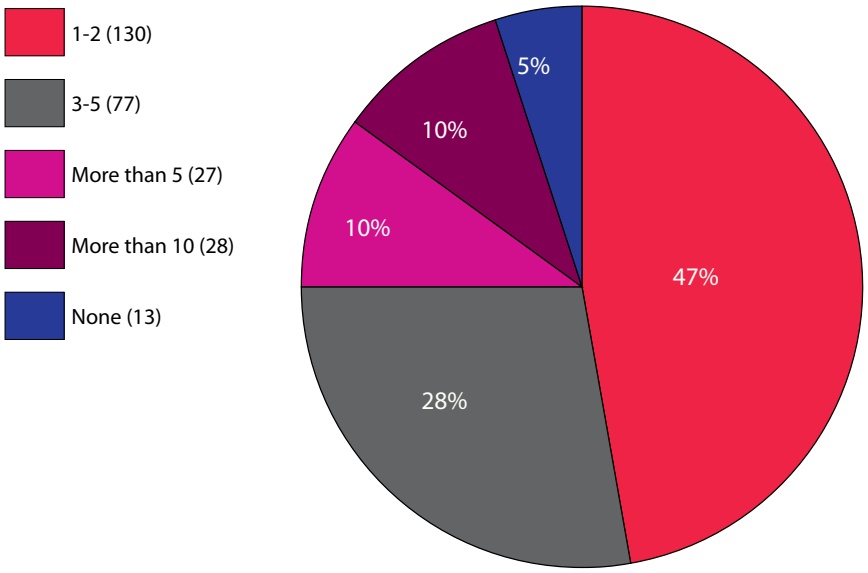
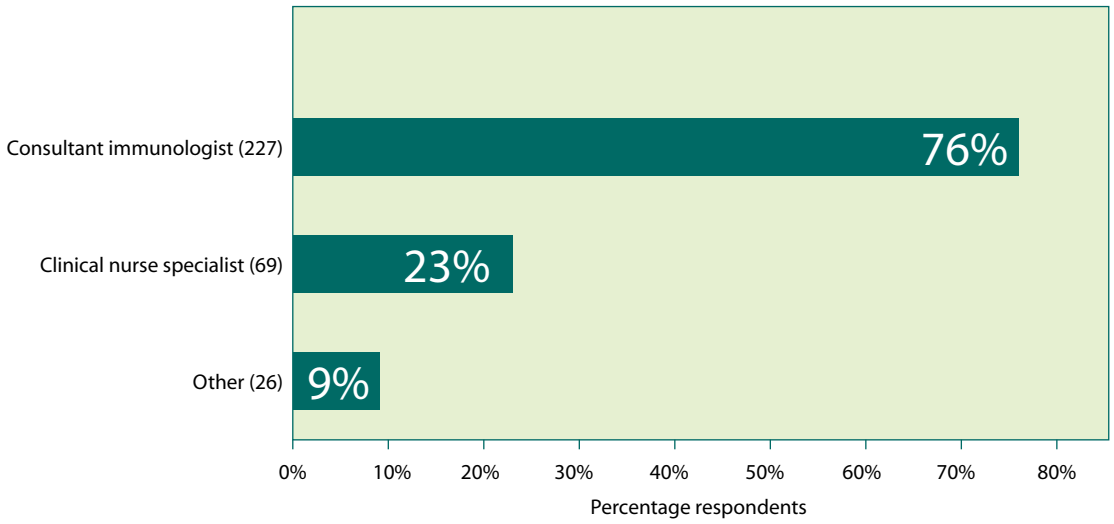


CHART 26: TYPE OF HEALTHCARE PROFESSIONAL INVOLVED IN THE RESPONDENTS FOLLOW-UP



47% (130) of respondents attended 1-2 regular outpatient clinics for assessment and follow-up (including monitoring of antibody levels, blood counts and liver function tests) in the past 12 months relating to the management of their PID condition (chart 25), with 76% (227) reporting that the follow up was carried out via the consultant immunologist *(chart 26)*.

CHART 27: TYPE OF HEALTHCARE PROFESSIONAL INVOLVED IN THE RESPONDENTS FOLLOW-UP. "OTHER" CATEGORY EXPLAINED (N=236)

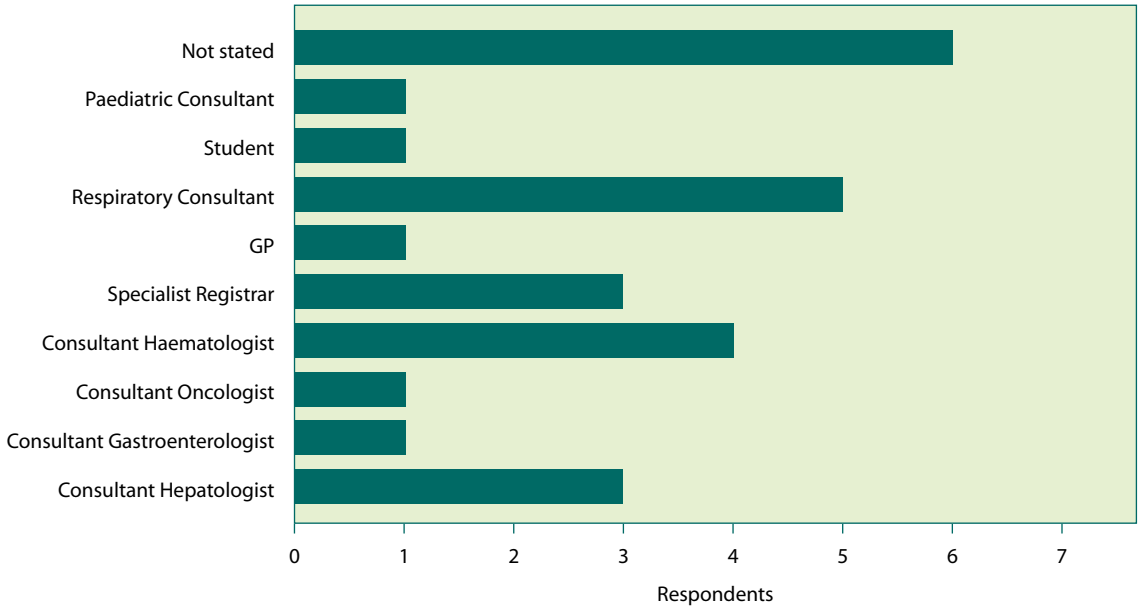
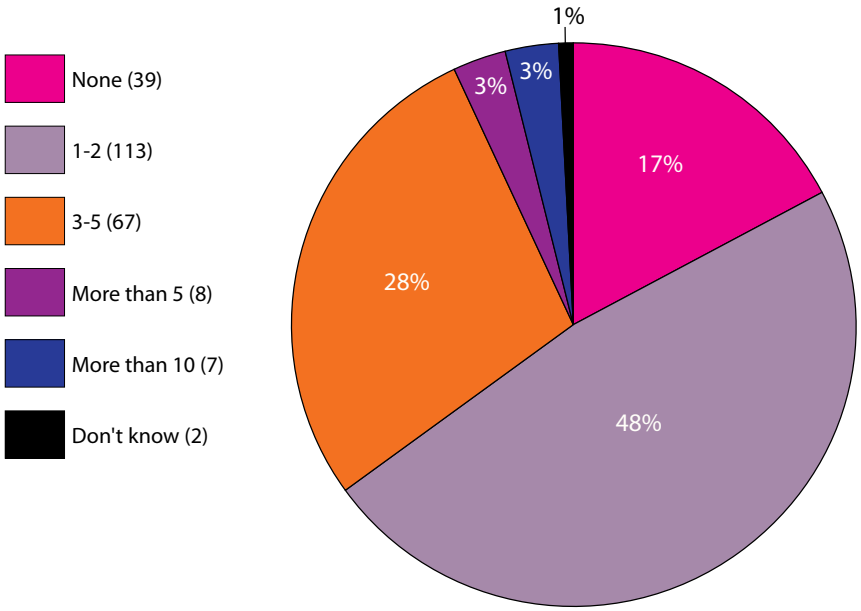


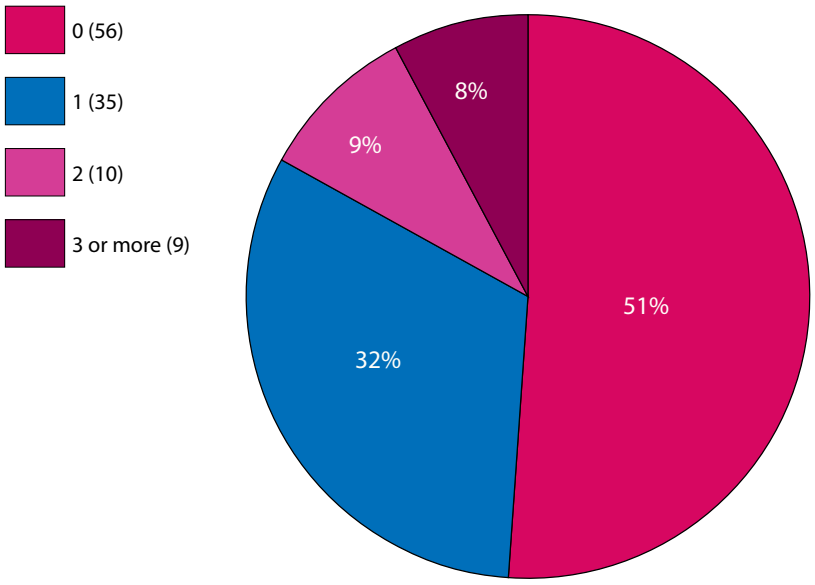
CHART 28: NUMBER OF OTHER SPECIALISTS RESPONDENTS HAD APPOINTMENTS WITH AS A RESULT OF THEIR PID (N=236)



48% (113) of respondents saw 1-2 additional specialists as a result of their PID, however this could range between 0-10 or more additional appointments per patient *(chart 28 and appendix 1: chart q).*

40% (47) of respondents had at least one separate inpatient stay within the past 12 months relating to the management of their PID. The overall response varied from 0-5+ inpatient stays. *(appendix 1: chart r).*

CHART 29: NUMBER OF INPATIENT STAYS THAT WERE REPORTED AS EMERGENCY OR UNPLANNED (N=110)

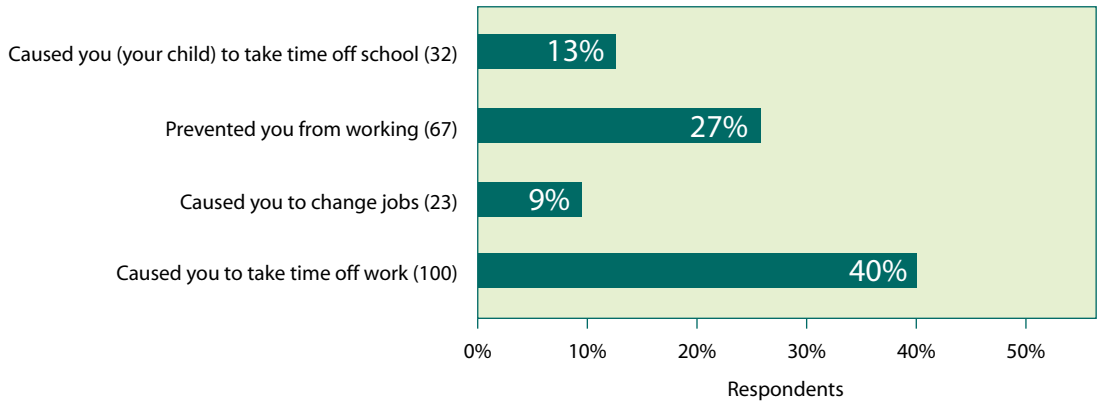


Of concern is the fact that almost half of the inpatient stays were emergency or unplanned, which add additional costs to the NHS and adds further burden to already stretched A&E services *(chart 29)*.

7. Quality of life

Being diagnosed with PID and on long-term medical treatment can have a significant impact on a person's quality of life. 40% (100) of respondents reported that their condition and taking immunoglobulin replacement therapy had caused them to take time off work and more alarmingly 27% (67) of respondents claimed that it prevented them from working all together. For children living with PID 13% (32) of respondents claimed that it caused them to take time off school. *(chart 30)*.

CHART 30: PERCENTAGE OF RESPONDENTS WHO REPORTED THAT THEIR CONDITION AND TREATMENT REGIME AFFECTED THEIR EDUCATION AND OCCUPATION (N=248)



These results indicate that it is essential that clinicians and patients/carers collectively decide on the ideal treatment regimen taking into consideration lifestyle, employment and educational needs. The impact otherwise could lead to consequences across a spectrum of a patient's life as this survey suggested:

Recreation

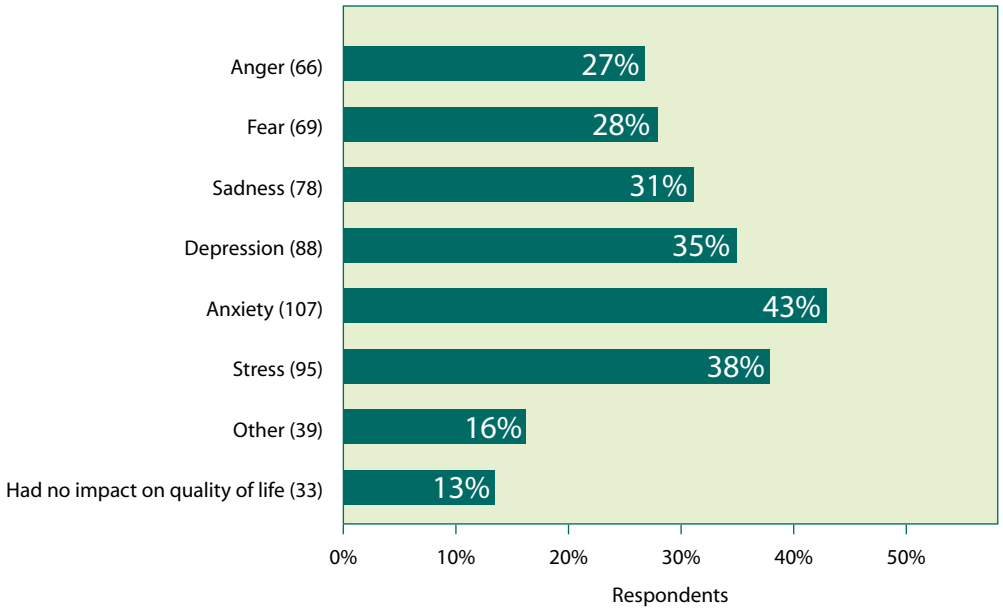
- 23% (57) of respondents felt limited on their ability to travel/enjoy holidays.
- 18% (45) of respondents felt limited on their ability to socialise.
- 26% (64) of respondents claimed they had difficulties managing household activities.

Relationships

- 29% (71) of respondents felt their condition affected their relationships with friends.
- 23% (57) of respondents felt their condition affected their relationship with their spouse or significant other.
- 18% (45) of respondents felt their condition affected them having and raising children.
- 26% (64) of respondents felt their condition affected their relationships with parents siblings and other relatives.

In addition, 215 (out of 248) respondents reported that their condition and treatment regime had an emotional impact on their wellbeing:

CHART 31: PERCENTAGE OF RESPONDENTS WHO REPORTED THAT THEIR CONDITION AND TREATMENT REGIME AFFECTED THEIR EMOTIONAL WELLBEING (N=248)



> *"I live alone. Enough said"*
>
> *Patient*

> *"Really changed what kind of life I live"*
>
> *Patient*

> *"Suicidal"*
>
> *Patient*

> *"Fatigue is so more emotional"*
>
> *Patient*

> *"Long term impact is of concern"*
>
> *Patient*

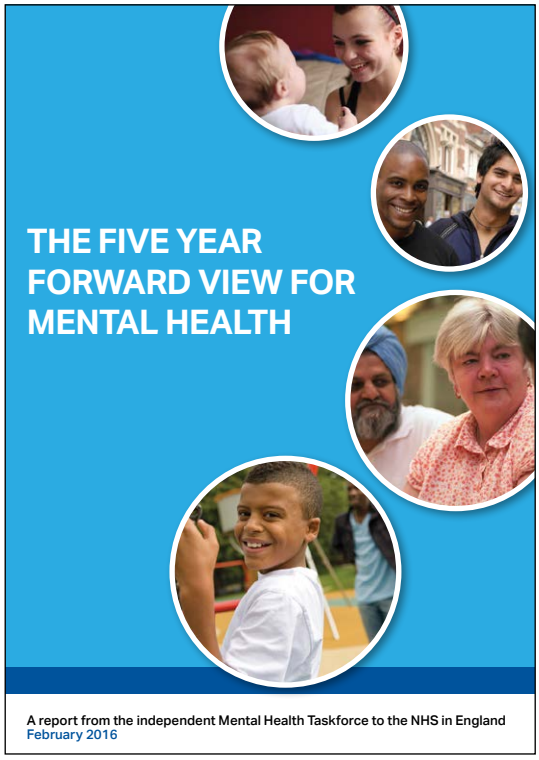
> *"Regret about my general ill health"*
>
> *Patient*

> *"Anger that there isn't a lot of awareness about PID. People think because you look OK, there is nothing wrong!"*
>
> *Patient*

> *"My child self harms"*
>
> *Patient*

Patients have needs other than the treatment of their specific health conditions. There should be recognition of the potential need for psychological and emotional support, as well as of the importance of meeting fundamental needs such as nutrition and pain management. In 2011, the Coalition government published a mental health strategy[31] setting six objectives, including improvement in the outcomes, physical health and experience of care of people with mental health problems, and a reduction in avoidable harm and stigma. February 2016 saw the publication of *The Five Year Forward View for Mental Health*[32], which calls for a fresh mindset and seeks strong leadership to tackle unwarranted variation in mental healthcare quality and outcomes.

Only 25% (63) of respondents have access to additional specialist services or health care professionals to ensure their quality of life is maintained e.g. psychological support, social care, however 65% (167) are content with their quality of life right now.



> *"The diagnosis and treatment have improved most aspects of my daily life. I am so much healthier and in control of my health"*
>
> *Patient*

> *"Diagnosis has improved my quality of life as correct treatment administered and constant infections controlled"*
>
> *Patient*

> *"I have dealt with it the best I can"*
>
> *Patient*

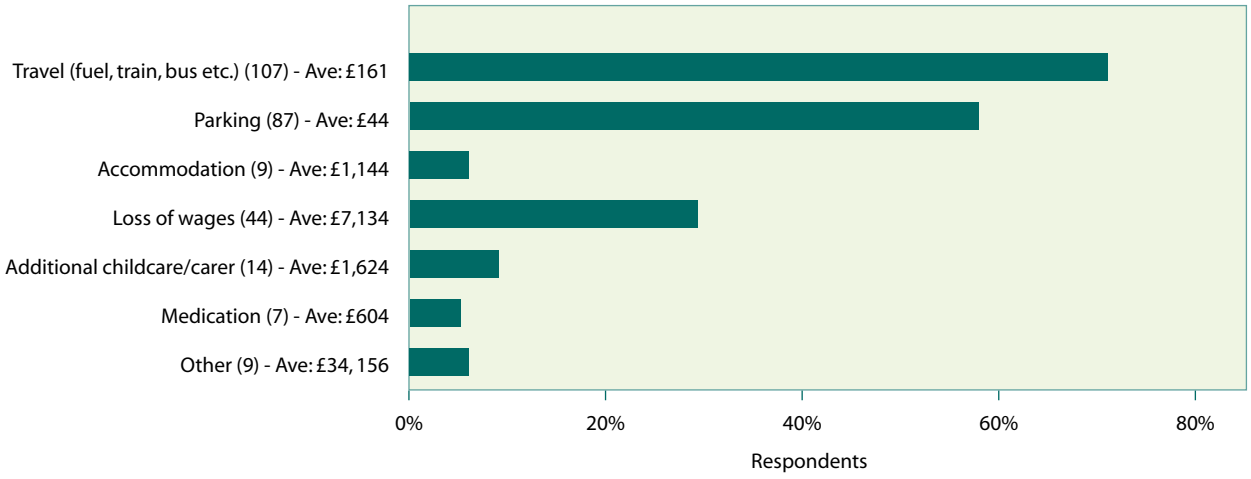
8. FINANCIAL BURDEN

Respondents were asked about the financial burden imposed on them due to their condition, taking into account factors, such as their domestic, social and work situation and their previous experience of healthcare, that may:

- Impact on their health condition.
- Affect their ability or willingness to engage with healthcare services.
- Affect their ability to manage their own care and make decisions about self-management and lifestyle choices.

59% (151) of respondents faced extra expenses in the process of receiving immunoglobulin replacement therapy. The most common expense incurred by 71% (107) of respondents was for travel costs, averaging an annual expenditure of £161. The largest expense incurred was for loss of wages, averaging £7,143 per annum amongst 29% (44) of respondents. 7 patients specifically reported an average cost of £604 per year on additional medications.

CHART 32: TYPE OF FINANCIAL EXPENSES RESPONDENTS AND THEIR FAMILY INCURRED ON AN AVERAGE 12 MONTHS (N=151)

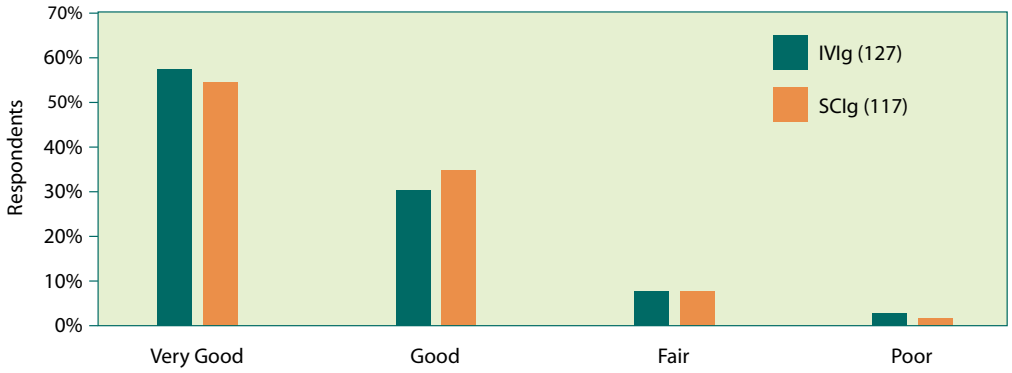


The majority of respondents (91%, n=219) were not aware of any financial support available from charities and social services.

9. OVERALL EXPERIENCE

The majority of respondents rated their overall experience with regards to the treatment and care they have received for managing their PID condition as either very good (52%) or good (33%), but with 15% of respondents rating their overall experience as either fair or poor, there is a greater need to improve service in PID care. *(appendix 1:chart s).*

CHART 33: OVERALL EXPERIENCE REPORTED BY RESPONDENTS VIA TYPE OF TREATMENT ADMINISTRATION (IVIG N=127) (SCIG N=117)



There was no significant difference between the overall experience of respondents on IVIg or SCIg.

Data limitations

There are a few limitations that should be considered when interpreting this survey data.

- Just over half of all the respondents that completed the survey were aged between 35-49 (29%) and 50-64 (30%), and therefore is not representative of all age groups. When compared to the UK PIN registry data it is the younger population that is not proportionally represented.

UKPIN registry (accessed October 2017)[36]		PID UK survey sample	
Under 16	16%	Under 16	7%
16-39	32%	16-24	4%
40-59	27%	25-34	10%
Over 60	25%	35-49	29%
		50-64	30%
		65+	20%

- Parents and carers of children and older people are only represented in this survey as a small proportion 17%.
- The percentage split between IVIg and SCIg was representative of the UKPIN registry, however these were not broken down via age groups, as per the report, so we are unable to compare.

UKPIN registry (accessed October 2017)[36]		PID UK survey sample	
IVIg	51%	IVIg	53%
SCIg	49%	SCIg	47%

- This is the first time, to our knowledge, that this type of survey has been conducted in the UK with this patient group therefore there is no baseline comparator of the patient experience over time.
- Not all respondents completed all the questions within the survey.
- In addition, recall failure, misunderstanding of the question, or both, can contribute to data inaccuracies. However, our survey reflects the current difficulties of everyday living and the impact of long-term treatment faced by patients living with PID.

Conclusion and proposed recommendations

The results of this survey highlight the importance of **early diagnosis** and providing **access to different treatment options** and **modes of administration** to ensure individual patient needs are best met. Our analysis and research findings show that where the NHS makes concerted efforts to drive change and improve quality, the positive outcomes are reflected in patients' feedback.

Based on the key areas that have emerged from this survey, we make the following recommendations:

> *"Once I was diagnosed I couldn't fault the treatment, nurses, consultant - all the team are excellent. I still have issues with my local surgery and the nurses are great at making sure I get the right medication, which relieves the stress from me having to explain my condition and need for antibiotics"*

Recommendation #1: Improve the knowledge and awareness amongst healthcare professionals in the diagnosis and management of PID. Patients often report an extended period of symptoms and repeated visits to their GP and specialist outpatient clinics before being referred to an immunologist and diagnosed with PID. This has a significant impact on a patient's quality of life and NHS resources as a consequence of managing the symptoms and complications resulting from their undiagnosed PID.

> *"It took many years to get a proper diagnosis, by which time I was unable to work as very ill"*

The impact of a late diagnosis of PID is too significant to ignore. There is clearly a need to raise awareness and understanding amongst healthcare professionals, particularly GPs to ensure that they are able to identify the signs and symptoms and make a referral to a specialist immunology centre.

> *"I wish more GPs were aware of PID before patients are diagnosed"*

> *"I am so appreciative of the treatment and support that I receive. I am fortunate that I have not been left with severe problems as a result of the lack of diagnosis. I feel that GPs should be made more aware of this area of medicine"*

Recommendation #2: Join-up services to improve the continuity of care for people living with PID. Continuity of care and co-ordination across organisational and professional boundaries is of prime importance in achieving good outcomes for patients with long-term conditions. Management continuity is relevant whenever a patient is receiving care from more than one clinician or provider. It concerns the processes involved in co-ordinating, integrating and personalising care in order to deliver a high- quality service[33]. Considering PID patients generally see a range of specialists and primary care providers during the course of their life, it is essential that all centres work to an agreed standard to ensure there is effective communication and integration of care and patients feel empowered and informed to be actively involved in their care plan. The GPs clinical responsibility as coordinator of care for patients includes helping patients to understand and plan their treatment, navigate unfamiliar services successfully and remain engaged with their care. Where appropriate patients and carers should be signposted to patient organisations such as PID UK that can provide further advice and support with their ongoing needs.

> *"The problem has largely been ignorance of the GP and too many people managing my care"*

> *"The complexity of my GP surgery being able to keep abreast of my repeat prescriptions (which change frequently) means that often they are wrong and require another trip to sort them out. Poor connectivity between specialised tertiary services and secondary and primary care results in the PID patient often having to broker and construct their own care pathway often at a time when they have least energy to do it"*

Recommendation #3: Ensure patients are active participants in their treatment choices, taking into consideration lifestyle needs and patient convenience. It is important that patients are offered full choice of treatment and made aware of the available routes of administration. Most patients wish to have flexibility in the system of ongoing PID care so that at different times in their PID journey they can choose different approaches, such as:

- When necessary attending hospital regularly.
- At other times effectively undertaking self-care at home or in a community setting.
- Having access to a specialist nurse to discuss complications, concerns, and co-ordinate their care.

There is good evidence that immunoglobulin replacement therapy prolongs survival and reduces morbidity and that administration by either the intravenous or subcutaneous route contributes to an improvement in QoL[34]. Furthermore, while current immunoglobulin treatments were generally associated with high levels of satisfaction, differences were seen with regard to patient experience and acceptability of certain convenience aspects of immunoglobulin treatment between patients treated with IVIg and those treated with SCIg. Utilising homecare for immunoglobulin treatment is vital to deliver the greatest possible flexibility for patients and optimise patients' control over their condition and therefore should be offered to all appropriate patients as a treatment option.

Recommendation #4: Expand nationally recognised performance metrics and actively performance monitor existing measures for PID to improve outcomes that matter to patients. The impact of living with PID for a person can be significant economically as well as socially. Untreated or inadequately treated syndromes can result in severe infections requiring hospitalisations, lost days from work or school, emotional distress, and fewer social interactions[35]. The cost of treatment includes but may not be limited to:

- Days missed from school or work.
- Hospital stays.
- Antimicrobial therapies and other treatments.
- Diagnostics and ongoing disease management.
- Management of complications such as autoimmune disorders.
- Treatment of adverse effects.
- Rapid access back into clinic if required.

> *"I believe there should be more Government support for people with invisible illnesses, especially when it comes to work. Its almost impossible for people with immune disorders to maintain a good attendance record in work and therefore hard to find new employment as chances of making it past your probation period without illness is slim. It can also be incredibly hard in a work environment as time off can lead to being ostracized by colleagues who believe you "look healthy so are faking". There are also some days when your body is so tired and it hurts just to move around yet its not recognised as a disability*"*

* PID is covered under the Disability Act

The impact of PID on health and social care is not widely understood or accepted and therefore lacks prioritisation and investment both nationally and locally. At a national level NHS England through its relevant Clinical Reference Group (CRG) and NHS Digital should look to expand and develop performance metrics that support additional improvement in clinical and social outcomes for patients, such as:

- Time taken to diagnosis.
- Review of patient's education and employment status.

It should also ensure that providers are fully implementing the national service specification[28] (B09/S/a) specifically:

- The provider shall undertake standardised Patient Related Experience Measures (PREM) surveys for patients and carers on an annual basis and achieve >75% satisfaction and act on any deficiencies identified.
- The provider shall comply with the requirements of the Immunoglobulin Demand Management Plan.
- Patients should be offered a choice of route (intravenous or subcutaneous) and location (hospital or home) for immunoglobulin replacement therapy if appropriate. All patients should have the opportunity to be assessed for home therapy if appropriate and undergo competency testing at recommended intervals.
- Clinical immunologists should review patients regularly on an outpatient or daycase basis in order to detect and treat disease progression or onset of complications, assess possible prognostic factors and carry out regular risk assessments for continuing treatment with immunoglobulin or other therapeutic agents.
- Working actively towards the RCP QPID accreditation.

Recommendation #5: All PID patients must have access to additional specialist services or health care professionals to ensure their quality of life is maintained e.g. psychological support, social care. Psychological distress, depression and anxiety are common side effects of living with a lifelong chronic condition and receiving immunoglobulin treatment. Many PID patients feel isolated and unsure who they can turn to for support. Having the opportunity to meet with other people living with PID through support groups can offer an immense sense of support and relief that they are not alone. In addition, many PID patients and their families are dealing with a heavy financial burden as a result of their illness. It is important to provide financial and other social care related support (i.e. psychological services) available to patients and their families as part of the information and support included in their personalised care plan.

> *"This survey is a good idea and should help the NHS and ultimately others like me"*

Further research needs to be conducted to evaluate the quality of life (QoL) for people living with PID. NHS patient experience surveys can provide detailed data that serve many audiences and purposes. However, NHS data is currently underutilised, both locally for quality improvement purposes, and nationally to inform policy development as well as secondary research on the aggregated data, which could provide insights not observable at a local level. The NHS is more open than ever to hear patients' views, and their experiences with healthcare providers will help to improve care for all patients living with PID. We hope that this study will promote further use and understanding of this important condition.

APPENDIX 1

ABOUT YOU

CHART A: ARE YOU COMPLETING THIS QUESTIONNAIRE AS A: (N=301)

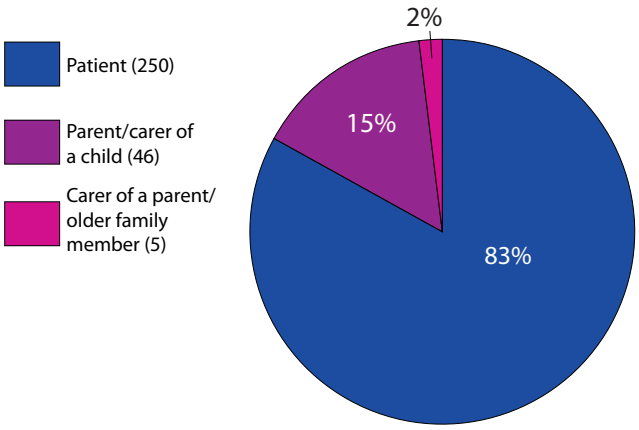
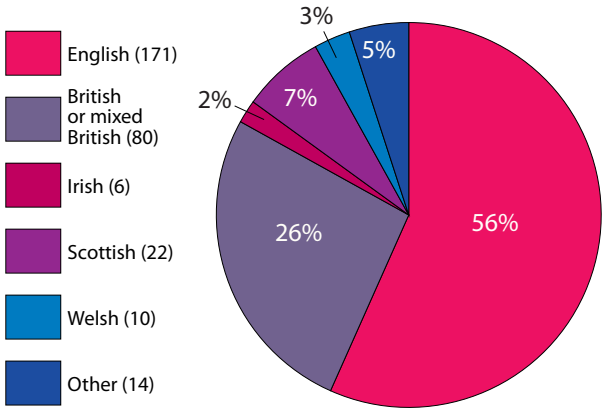


CHART B: THE NATIONALITY YOU MOST IDENTIFY WITH (N=303)



1. DIAGNOSIS AND ACCESS TO CARE

TABLE C: HOW DID YOU FEEL ABOUT THE AMOUNT OF TIME IT TOOK TO DIAGNOSE YOU?	
Negative	**Positive**
Extremely worrying for all the family.	I felt ok with the time.
I didn't know I had PID until a doctor treating me for something else sent me for a lot of blood tests, it was discovered by pure accident!	Diagnosis was fairly quick from blood tests at GP, then a couple of months until seen by a specialist. Steroids over the past 3 years made condition much worse.
It was very frustrating trying to get the referral from GP to a Respiratory Specialist. My sister was able to get an appointment quicker though I don't blame my GP for this.	Feel ok as it was done as a follow up to pneumococcal-associated HUS.
Felt like I was hitting my head against a brick wall.	Not applicable - blood test results referred to immunology dept.
Pretty disgusted - I thought I was a hypochondriac.	As this is a rare illness, nobody including our GP or at hospital A&E spotted the illness. It was fortunate that a junior doctor at the hospital spotted something that was more than just normal flu and lung infection.
Frustrated - I knew something wasn't right but the immune deficiency was only diagnosed after many other conditions had been ruled out.	Acceptable: as I worked in a farm environment other causes needed to be ruled out first.
Worried, there was no explanation for our daughter's symptoms and not knowing what her future prospects would be.	It was no ones fault. Was diagnosed while as an inpatient with unconnected symptoms.
As a parent, four years feels like a very long time, especially when my son was at the GP so often.	Very quickly.

As parents we spent 4 years trying to get help and be listened to until finally she nearly bled out due to bone marrow failure. We then had new consultants who are fantastic. I had called a meeting with the previous endocrine consultant 1 month prior to this episode and was told to expect this until she was 5! The previous consultant would not refer me onto anyone as he said "who do you want me to refer you onto? If I felt someone else needed to see her then I would refer her". He had also told our GP not to expect much from this child. Our GP refused to come to our house when she was poorly because they hadn't been told by our consultant that she was immuno compromised, because he wouldn't refer us to that specialist. So ultimately she was very poorly for a long time with no help and nearly died.	Although I was only formally diagnosed at age 42, I had been under the care of haematology services since age 18. Whilst I had not had access to regular IVIg until formal diagnosis, I did have reasonable antibiotic management, and management of auto-immune symptoms. I'm not that sure that formal diagnosis would have made huge differences to my day-to-day health at that time. Indeed, my haematologist had referred me to a clinical immunologist in my 20's who had assessed me and announced that 'I had nothing that he was interested in'. My guess is that I was being referred for research purposes (although not explained to me) and that a full service for people with CVID was not available at that time anyway.
When finally a respiratory consultant started to investigate I was referred to an immunology clinic, where I had 18 months of investigations.	I was lucky as diagnosed on first hospital stay for a mystery illness.
I would have been saved a great deal of illness if my GP had just completed a simple blood test.	Not unreasonable; I was lucky it was found at all.
I felt it took an eternity and at times I wanted to die.	It was too long but the tests to determine it are so specialised I can't see how it could have been faster.
It would have saved myself from having an operation on my sinus if I was diagnosed earlier and at times I was sent away from my GP thinking I was going mad.	Happy with the time taken.
Took quite a while and a lot of invasive surgeries.	Extremely cared for.
Horrendous! I saw more doctors and specialists than I can count and was doubted and not believed my whole life until in my mid-thirties. When I was 14 I asked my GP if there was something wrong with my immune system and he laughed at me.	Since various different problems /conditions were being investigated (including 1 other rare and another extremely rare diagnosis) I was pleased that a conclusion was eventually made that has helped me. Of course it would have been brilliant if this had been realised sooner, but every effort was made to work out what was going on with me.
Worried, as after many years of hospital visits and 3 - 6 monthly checkups I have been diagnosed with so many different conditions, from possible HIV to Lupus.	I was extremely fortunate to be diagnosed with a PID aged three in 1956.
Before being diagnosed my life was filled with frequent and prolonged infections many of which were quite debilitating and rather nasty. I'm not sure that I can adequately describe what I felt about it. It was the only life I knew. What I can say is for me being diagnosed (at the age of 24) was something of a eureka moment, something that could finally explain why I was the way I am. The fact that I was diagnosed so late has had a deep lasting effect on my life. So much time missed from school and always feeling so different from my peers, being severely bullied when I was at school because I was weak and sick. My outlook on life remains to be affected by this.	A little slow at first, but as soon as it was suspected the process was fast.
Initially I was quite cross because from a very early age I was always unwell, one infection after another, and I feel that had the information been out there, a lot of time and money could have been saved.	I understand that I was/am a complex case and that consultants tried their best to work out what was wrong with the resources they have. Obviously with better diagnostic tools and understanding of the disorder this would potentially speed up this process but I understand that as a rare and complicated disease this is very difficult.
Frustrated and upset - I didn't understand what was going on and the fact that medical professionals seemed to dismiss me, saying that I would get better didn't help - it made me question whether I was really ill, or whether I was just going crazy!	As symptoms and conditions did not feel too serious I always felt like I had minor health problems all my life. Only in the last 4 years have I been more prone to infections. New medicine and research has helped to diagnose me and help to improve my life slightly, which I am grateful for.
It should have been picked up much sooner. I was presenting at doctors with problems from a very early age. I was 36 when I found out something real was wrong with me!	Very satisfied. There was a history of the condition in my family so I was diagnosed at three months of age.

I had to go to a private BUPA hospital for diagnosis as the treatment I was getting on the NHS was erratic and no-one could make a positive diagnosis. I think the NHS should have done more to diagnose my condition but even though I had to pay to go privately I was just so relieved when I got a diagnosis and got appropriate treatment.	For 2/3 years I suffered chest infections including pneumonia before referral to Immunology Dept. I believe that at that time i.e. 1999 the average seemed to be in excess of 5 years so I was lucky and pleased.
I feel let down that it took so long for someone to diagnosis my daughters condition. She spent months and years unwell and due to late diagnosis, developed Bronchiectasis. We spent so many hours at GP surgeries, hospitals and eventually private hospitals until we had a diagnosis. One consultant said we needed to get to the bottom of her problems and we should see an immunologist. Finally.	One question to my GP 5 years ago "why I am I always getting these infections?" Blood tests revealed my IgG to be 0.39. After many consultations I reached the immunology dept. Antibody replacement treatment was started 4 years ago and now I have never been fitter in my life.
Sometimes I feel quite upset as I probably was seen and expressed concern regarding my son by about twenty different healthcare professionals ranging from GP, health visitor and nurse. In the first seven months of my son's life we were never offered any further tests. We were given a range of diagnosis eczema, cow's milk intolerance and possible depression, anxiety on my part. At no time when I expressed concern was I asked what I wanted for my son. My son's condition was only first picked up as I demanded a referral from GP to A&E when he was not well at eight months old. They ran a blood test and found that his platelets were very low thus starting a six-month road to diagnosis - originally they thought it was Idiopathic thrombocytopenic purpura. What I'm trying to say is why are not full blood counts run as standard when a patient has repeated unexplained problems, they can tell so much. My son was very ill from about five weeks old and it took until he was 8 months old for a full blood count. During those eight months he was seen 30 times by the health visitor weekly at baby clinic, 20 plus by GP, 5 times by paediatrician and 4 times by dermo nurse specialist and 1 time by dietician.	One week from being admitted to hospital, very pleased for such a rapid diagnosis.
Anxious, depressed, panicked. I felt the NHS were more interested in protecting their budget than conducting the requisite tests to determine what was wrong with me.	My GP was very quick to have my immune system tested after 18 months of continuous infections. I just thought I was having a rough year, but he called me in to say it seemed a very high amount of illness. After he referred me I was diagnosed in 3 months.
As I was a baby I was not aware of the time it took. My mum said after the time it took time to diagnose me I deteriorated so I feel something should be available to test at birth.	Diagnosed immediately once referred to Immunologist after GP blood tests.
I was very angry, as I had been having swelling attacks and the only advice given was to "rest and relax, and let swelling go down". I had also undergone unnecessary surgery on two occasions.	I was simply grateful that CVID was diagnosed as the cause of other long term issues - chest and sinus infections.
Although my GP made early IgA deficiency diagnosis there was great resistance and denial by hospital consultants to correct interpretation when COPD and repeat infections involved. Eventually a second hospital and an immunologist provided stability and correct antibiotic treatment. First hospital's medical director had to personally apologise to me for the mistreatment, although the consultants involved were not disciplined or apologetic. I recently moved to another area with a large immunology/allergy centre which has, after initial involvement, failed to provide ongoing support unless a diagnosed infection is found. Centre also refused to support the use of IVIg for related autoimmune disease requiring repeat appeal to NHS England and referral to approved clinical immunology centre.	Pleasantly surprised. I had experienced one minor facial swelling and a second major facial swelling a fortnight later-both treated as an allergy. On discharge after the second one, I was referred to an allergy clinic and attended 3 months later. I was told that it could not be an allergy. Blood samples revealed potential problems and further blood samples were taken 3 weeks later, when I was informed of the anticipated HAE problem. Formal diagnosis was confirmed 2 weeks later, less than 4 months after the major incident, with no additional attacks.

2. TREATMENT AND CHOICE

CHART D: DO YOU FEEL THAT YOUR LIFESTYLE, PERSONAL PREFERENCE, CULTURAL BELIEFS AND RIGHT TO CHOICE WERE RESPECTED WHEN DECIDING YOUR TREATMENT OPTIONS? (N=303)

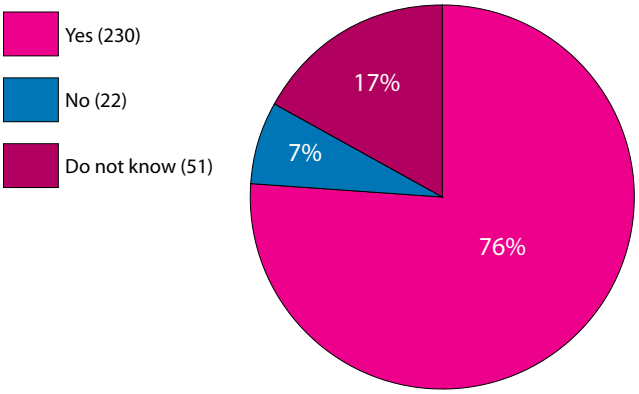


CHART E: IN RELATION TO YOUR IMMUNOGLOBULIN REPLACEMENT THERAPY, HOW DO YOU CURRENTLY RECEIVE IT? (N=225)

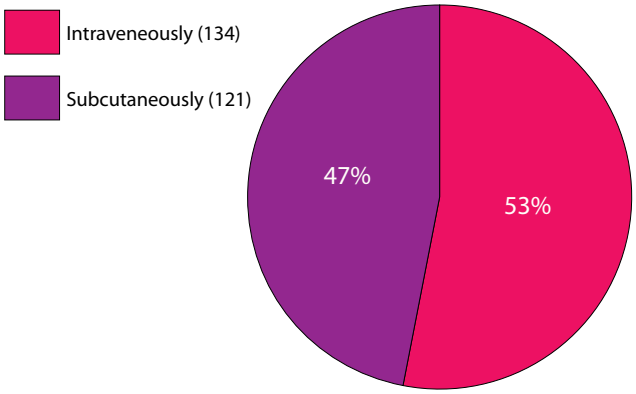


CHART F: WERE YOU OFFERED THE CHOICE OF WHERE YOU WANTED TO RECEIVE YOUR IMMUNOGLOBULIN TREATMENT. I.E. AT HOSPITAL OR HOME? (N=257)

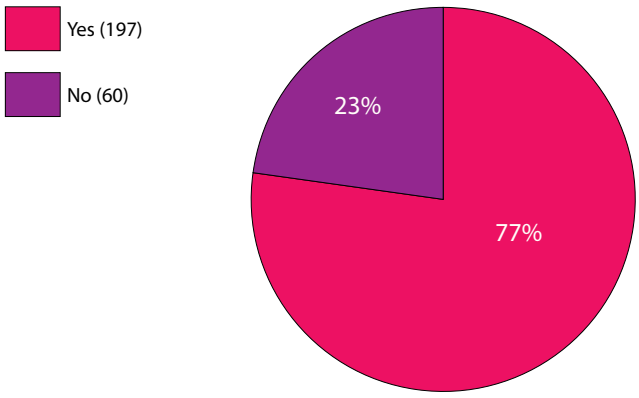


CHART G: ARE YOU SATISFIED THAT YOU RECEIVE YOUR IMMUNOGLOBULIN THERAPY IN AN APPROPRIATE LOCATION. E.G. AT HOSPITAL OR HOME? (N=255)

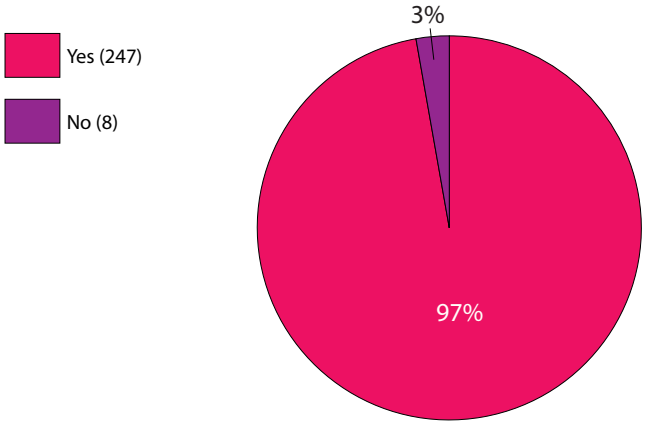


CHART H: HOW FREQUENTLY DO YOU OR YOUR HEALTHCARE PROFESSIONAL TAKE BLOOD SAMPLES AS PART OF MANAGING YOUR TREATMENT REGIME? (N=265)

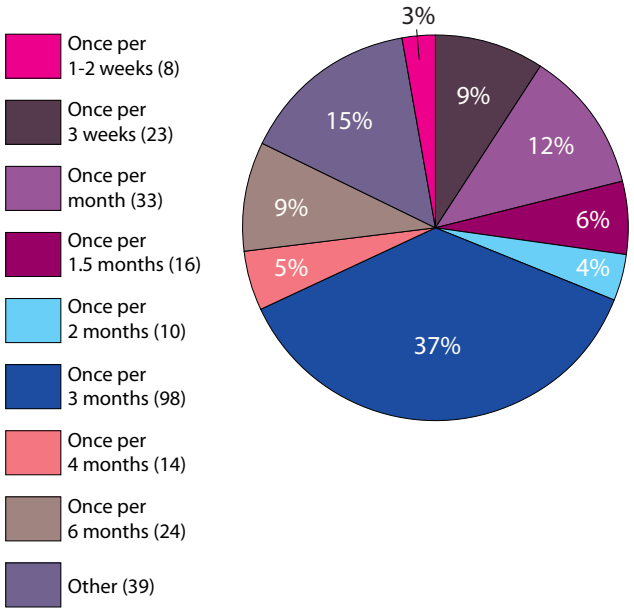
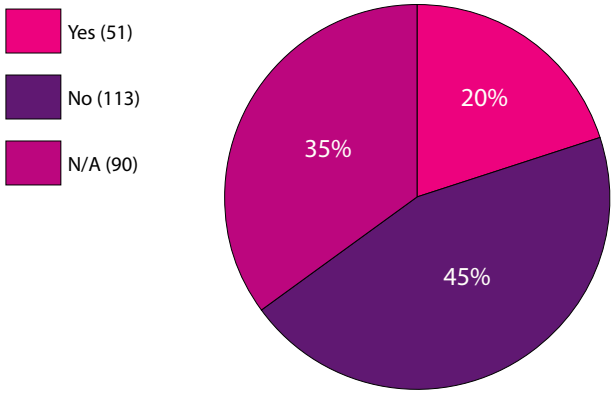


CHART I: DO YOU HAVE TO TAKE ANNUAL LEAVE TO ATTEND APPOINTMENTS? (N=254)



3. HOMECARE TREATMENT

CHART J: HAS YOUR ABILITY TO SELF-ADMINISTER BEEN TESTED AFTER RECEIVING HOME THERAPY TRAINING? (N=131)

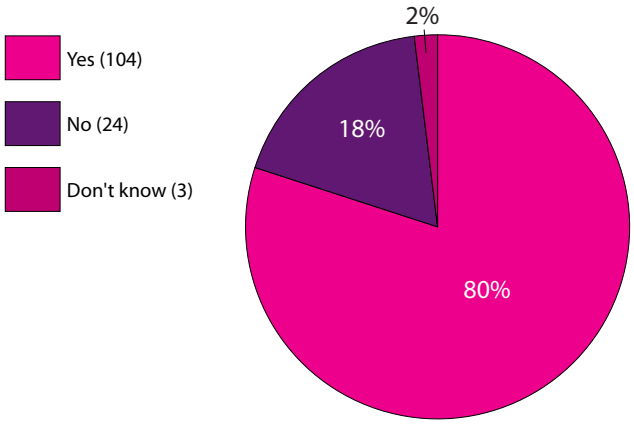


TABLE K: DO YOU HAVE ANY CONCERNS IN CONNECTION WITH THE EQUIPMENT AND/OR CONSUMABLES DELIVERED TO YOUR HOME?	
1.	Consumables have to be collected from local clinic.
2.	Poor condition of packaging by company concerned, vague delivery dates and times. This is common throughout all companies I have had dealings with.
3.	Not the most reliable home delivery company that I have been with.
4.	Deliveries not always accurate.
5.	Was given very fast tubing and had to ask for slower, that took several emails and phone calls to achieve.
6.	Have had a couple of items faulty.
7.	1 in every 3 orders has something wrong, requiring me to chase them and get correct equipment sent or return equipment I don't need.
8.	I have to collect all my equipment from three different locations, none of which are within walking distance and I don't drive. Obviously when you are regularly sick this can mean it is difficult to travel on public transportation which is full of germs and it is then hard to carry large loads home again.
9.	It can be a bit of a gamble whether or not I get the correct items.
10.	There are occasional issues with the homecare company who deliver e.g. wrong or forgotten consumables.
11.	I order from district nurse but due to staff change over often misunderstand requirements.
12.	The provider does not always respond exactly to my equipment and consumable requirements.
13.	One of the plugs is faulty.
14.	Brought pump myself. Getting the consumables paid by the medical aid is also a hassle.
15.	Sometimes the frequency is too often and I have surplus consumables. Full sharps bin not always collected by driver.
16.	The delivery was quite difficult as at first only part of the equipment had been provided leading to booking a last minute hospital session. Then it was not for the correct number of months. Finally, the needles bin has never been collected after 7 months now, therefore, I feel the company who provides the medicine and the equipment could be a bit better organised.
17.	Just that it will all be delivered on time.
18.	The paper 'op-towels' to put equipment on has tripled in size since getting it delivered to home. Could probably cover a snooker table with it! Just seems a bit of a waste on paper for resting a needle on. Not a big issue I know! Also, I now have enough medi-swabs/air inlet needles/plasters to last a life time. Everything else is all good!
19.	Batteries get used up a lot off the time and have quite a lot of occlusions.
20.	It has been difficult to get sharps bins collected once full but I think this has been sorted out after many years.
21.	There are sometimes items missing from my delivery, but the service has improved.
22.	Timing of delivery during the day to make sure someone is available. However, my parents/partents-in-law are always willing to come to the house as both myself and my wife work during the day.
23.	Would be nice to have a pump to administer the immunoglobulins.
24.	My pumps are not working very well.
25.	Sometimes I'm not informed the consumables have arrived at the GP surgery or they forget to give me a packet and I have to chase them up.
26.	Items regularly wrong or short.
27.	They won't give a time slot.
28.	I'm not keen on the new butterfly needles I've been sent recently. The connection to the syringe seems more difficult. Also I don't like the tiny sharps bins they're delivering now.
29.	First time I was delivered the wrong syringes.

CHART L: IS YOUR HOME CARE TREATMENT REGULARLY REVIEWED? (N=131)

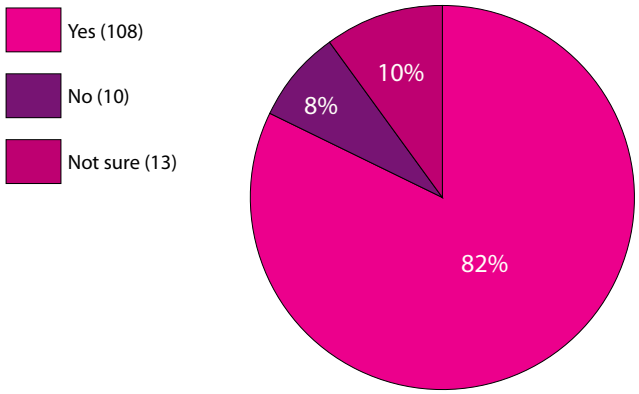
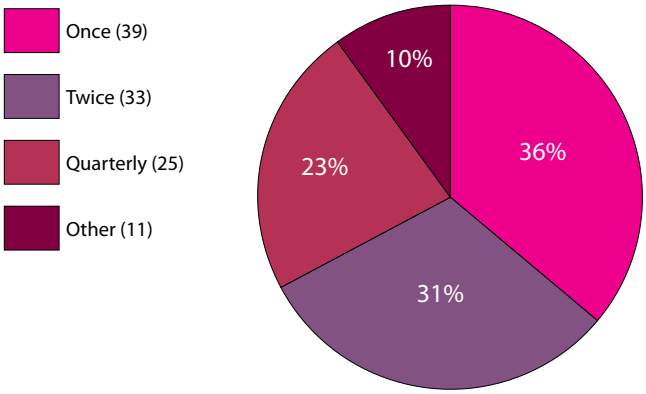
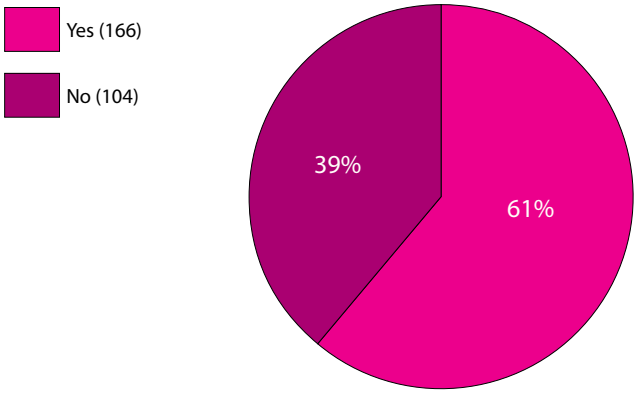


CHART M: IF YES, HOW OFTEN IS YOUR HOMECARE TREATMENT REVIEWED IN A YEAR? (N=108)



4. INFORMATION, COMMUNICATION AND EDUCATION

CHART N: WERE YOU MADE AWARE BY YOUR HEALTHCARE PROFESSIONAL AS TO THE IMPACT ON EVERYDAY ACTIVITIES YOU WOULD UNDERGO LIVING WITH PID? (N=270)



5. SUPPORT

CHART O: WERE YOU AWARE OF AVAILABLE SUPPORT TO HELP YOU OVERCOME ANY FEARS OR ANXIETY RELATING TO YOUR PID CONDITION AND TREATMENT THAT YOU MAY EXPERIENCE? (N=277)

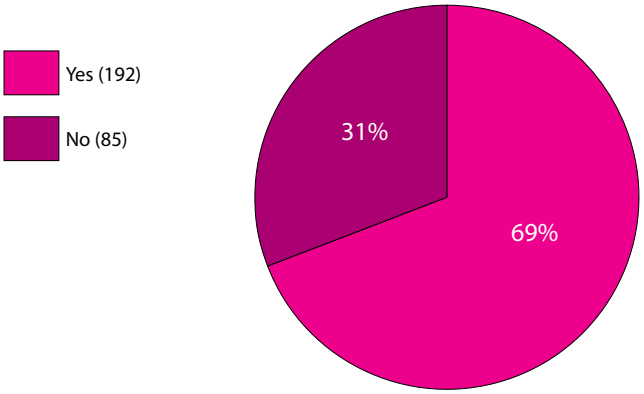
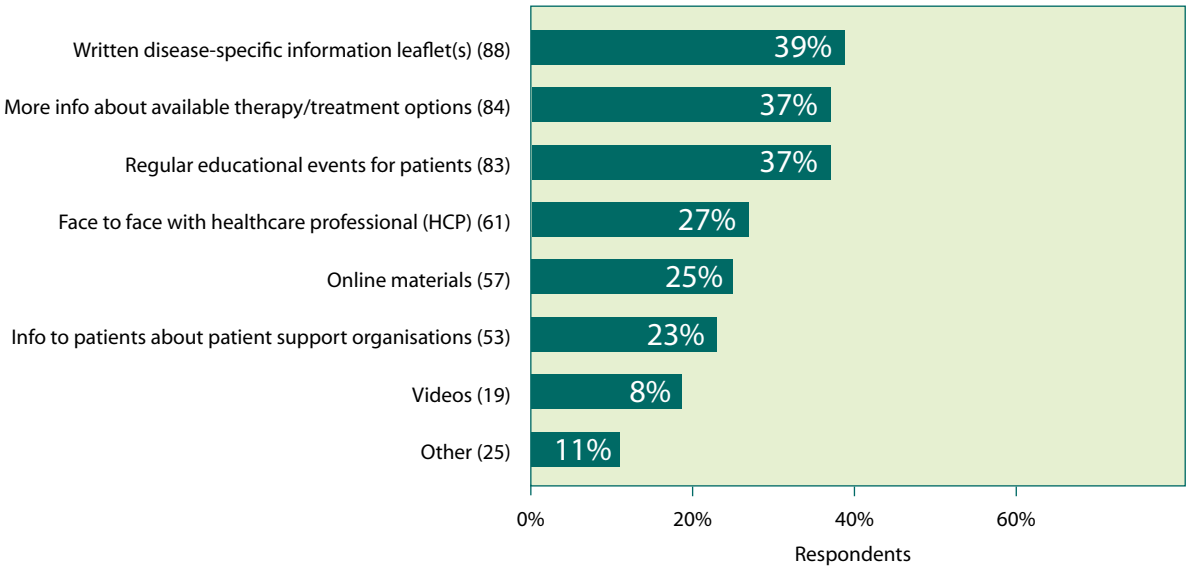


CHART P: WHAT WOULD HELP YOU TO MANAGE YOUR CONDITION BETTER? (N=226)



6. LONG-TERM MANAGEMENT

CHART Q: PLEASE STATE HOW MANY ADDITIONAL APPOINTMENTS YOU ATTENDED IN THE PAST 12 MONTHS WITH OTHER SPECIALISTS. (N=243)

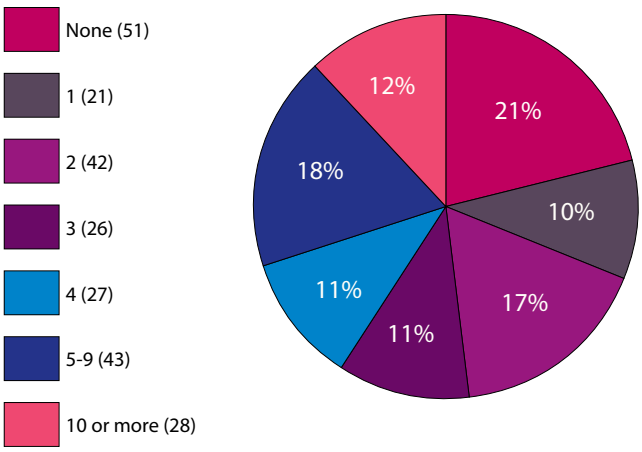
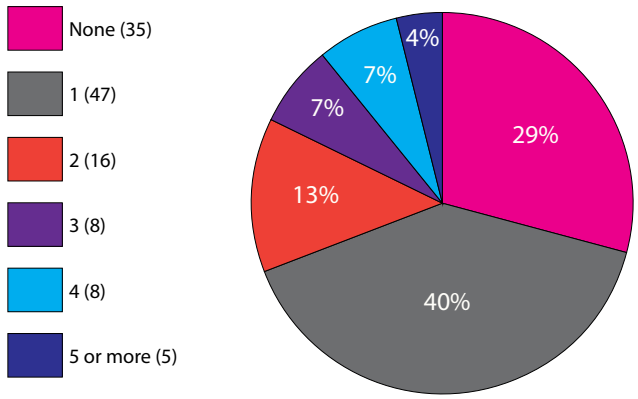


CHART R: HOW MANY SEPARATE INPATIENT STAYS HAVE YOU HAD IN THE PAST 12 MONTHS RELATING TO THE MANAGEMENT OF YOUR PID CONDITION? (N=119)

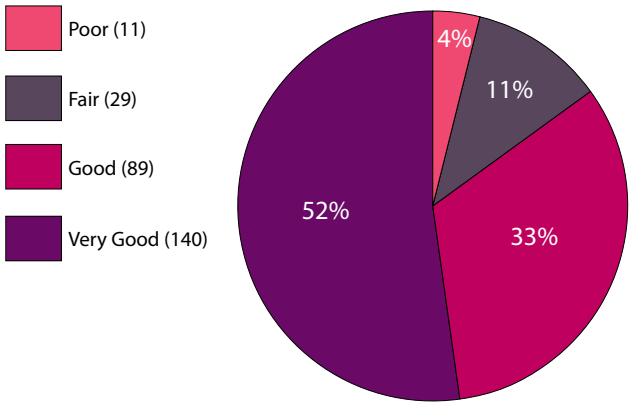


On average - 1.3 stays

Sections 7 (quality of life) and section 8 (financial burden) are refered to in the main body of the report.

9. OVERALL EXPERIENCE

CHART S: HOW WOULD YOU RATE YOUR OVERALL EXPERIENCE WITH REGARDS TO THE TREATMENT AND CARE YOU HAVE RECEIVED FOR MANAGING YOUR PID CONDITION? (N-269)



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