IMMUNOGLOBULIN REPLACEMENT THERAPY FOR PRIMARY ANTIBODY DEFICIENCY MODEL OF CARE

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IMMUNOGLOBULIN REPLACEMENT THERAPY MODEL OF CARE

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Executive Summary

Antibody deficiencies are a group of Rare Diseases which are the most common subgroup of Primary Immunodeficiencies. The maximum number of patients affected by this condition is 1:10,000 of the population. The only treatment for significant antibody deficiency is immunoglobulin replacement therapy (IgRT), which is required on a lifelong basis. When diagnosed and treated promptly, before the onset of complications, patients with antibody deficiencies enjoy a near-normal life-expectancy. When complications are present at diagnosis, or treatment is inadequate, patient survival is impaired, and patients are at risk of sepsis, as well as the development of a chronic lung disease called bronchiectasis (similar to the lung disease seen in Cystic Fibrosis). This is a preventable form of disability.

Immunoglobulin replacement therapy may be given intravenously, subcutaneously, either pump-driven or by rapid push, or by facilitated subcutaneous infusion. The goal of immunoglobulin therapy is to prevent infections, and restore an adequate IgG level. Once this is achieved, the route of immunoglobulin administration does not affect treatment outcomes. Treatment may be administered in hospital, however this requires 3 weekly infusions and disrupts education, working and family life. For suitable patients, following training, home therapy has been shown to be highly effective, associated with improved Quality of Life, and significantly decreases healthcare costs. In July 2017, there were 270 patients (223 adults and 47 children) receiving IgRT for antibody deficiencies. Of these, 124 (46%) had infusions in the immunology centre, 36 (13%) in local hospitals and 110 (41%) were undertaking home therapy. Hence there are currently a total of 270 patients in Ireland, who are included in the scope of this Model of Care.

Ireland is the only country in Western Europe where there are still limitations in accessing subcutaneous immunoglobulin for suitable patients. This forces immunologists to continue to treat patients with more expensive, hospital based infusions, with negative consequences for patients' quality of life. Rates of home therapy provision among the 3 adult immunology centres were between 18% and 60%, while 68% of paediatric patients have access to home therapy. A review of Irish patients undertaking home therapy has shown very high levels of safety and patient satisfaction.

It is recommended that:

- When the decision is taken that IgRT is required, this should be started within 4 weeks, and no later than 8 weeks
- All patients who are suitable for home therapy should be offered training, and training should commence within 8 weeks

- Patients should have the right to choose the route of administration which is least burdensome, from medically appropriate options.
- Administrative obstacles to home therapy should be addressed
- The governance of patients undertaking IgRT at home remains with their immunology centre. Appropriate support and review processes must be in place.

The recommended Model of Care aligns with the fundamental principles of Slaintecare, and advocates treatment at home for all suitable patients, and as close to home as possible where home therapy is not an option.

1. Introduction

1.1 Background

The primary function of the immune system is to protect us from infection. The immune system is divided into the innate immune system (a rapid response system) and the adaptive immune system. The adaptive immune system learns and improves from previous exposure to infection or vaccines, and produces a slower, highly specific and effective response. The adaptive immune system is further divided into the antibody-mediated response, which provides protection against bacterial infection, and the cellular response which plays the primary role in defending against viral, fungal and mycobacterial infection. Antibodies are the proteins which make up our immunoglobulins.

Patients who have an impaired ability to make antibodies experience recurrent bacterial infections. Typically, infections affect upper and lower respiratory tract, but may affect any part of the body, and severe invasive infections such as meningitis, osteomyelitis and sepsis occur in some patients. Delayed diagnosis allows recurrent episodes of chest infection to cause permanent damage to the lungs (bronchiectasis), where the airways become dilated and irregular. This is a preventable cause of chronic disease and disability. Hence early diagnosis and effective management of immunodeficiency should be available to all patients. The mainstay of treatment for patients with significant antibody deficiencies is immunoglobulin replacement therapy (IgRT).[1,2] The primary goal of immunoglobulin replacement is the prevention of sepsis, pneumonia and other serious acute bacterial infections, as well as reducing recurrent sinopulmonary infections and improving quality of life. Immunoglobulin products for replacement therapy are on the WHO Lists of Essential Medicines for adults and children (WHO 2015).[3]

Primary antibody deficiencies may be due to a known genetic mutation or be of unknown cause. Antibody deficiency may also be secondary to known causes, such as leukaemias and lymphomas, following chemotherapy or treatment for autoimmune disease and following transplantation (termed secondary antibody deficiencies). Typically, patients with primary antibody deficiencies are managed by clinical immunology teams. The most common group of patients with secondary antibody deficiencies (associated with lymphoproliferative diseases) are managed by haematology teams, who manage both the primary haematological disease as well as the complicating antibody deficiency. Clinical immunology

teams are involved in managing a small number of complex patients with secondary immunodeficiencies.

Immunoglobulin can also be used in high doses to modulate or adjust the immune response in some autoimmune disease. The doses used for immunomodulation are usually 5 times higher than replacement doses. Because of the high dose required in autoimmune disease, intravenous treatment is usually preferred, and many of the treatment options available to immunodeficient patients are not relevant. It is outside the scope of this Model of Care to consider immunomodulatory use of immunoglobulin, which is commonly used in neurology for management of chronic immune-mediated neuropathies, in haematology for management of autoimmune cytopenias, and occasionally in rheumatology.

1.2 Scope

The scope of this document is the management of IgRT in patients with primary immunodeficiency (PID), as there is international consensus on management of these disorders.

To prevent variation in access to treatment by cause of immunodeficiency within the same infusion unit, it is proposed that similar administrative arrangements are available to the small number of patients with complex secondary immunodeficiency managed within immunology units.

It is anticipated that a further model of care will be developed for the broader population of patients with secondary immunodeficiency. This will reflect currently evolving international guidance and consensus, and will acknowledge similarities (risk of serious infection, sepsis and long-term end-organ damage), but also differences (such as threshold for treatment and monitoring for recovery), between primary and secondary immunodeficiencies.

An additional Model of Care addressing the use of immunoglobulin for immunomodulation would also be required, as the type of immunoglobulin treatment for these patients is different to the approach used in immunodeficiency, and non-immunoglobulin options are also quite different.

1.3 Epidemiology

There is limited population based data on the number of patients requiring IgRT for the management of PID. It is estimated that a maximum of 1:10,000 of the population may require IgRT for management of a PID, however the number may be considerably lower than this.

PID is incorporated into the European Reference Network for Rare Immunodeficiencies, Autoinflammatory and Autoimmune Disease (ERN-RITA). The European Society for Immunodeficiency (ESID) is one of the associated Scientific Societies.

The UK Primary Immunodeficiency Network (UK PIN) developed a National Registry of patients with PID, which links to the European Society for Immunodeficiency (ESID) Database. A review of activity of the UK Registry from between 2008 and 2012 reported 36 of the 38 immunology centres in the UK engaged with the process, but only 71% had commenced entering data (Edgar et al; 2014).[4] Of the estimated 4000-5000 patients with PID in the UK at the time, 2,229 had been entered on the database. Patients with antibody deficiencies formed the largest group, with 1,358 receiving IgRT, of whom 1/3 were being treated in the home setting. The proportion currently having home therapy has increased to approximately 50% (Edgar JD; personal communication 2017). In Northern Ireland (population 1.811 million), in March 2017 there were 161 patients receiving IgRT in immunology centres – this equates to 1:11,248 of the population (Edgar JD; personal communication 2017)

It is notable that rates of primary immunodeficiency in the Republic of Ireland reported to the ESID database are approximately 50% of those in Northern Ireland [4] (Edgar JD personal communication 2017). Of note however, only one Irish centre is currently reporting to the Registry, with other centres going through Research Ethics Committee approval and/or identifying the necessary resources to contribute to this European initiative. Improved education for primary care and frontline physicians together with exploration of novel screening approaches are required to promote timely diagnosis of immunodeficiency. Such innovations, pioneered in other countries prevent medium term morbidity and long term cost. [5]

A telephone-based population survey to determine the prevalence of immunodeficiency has been performed in the United States, by an immunodeficiency advocacy group.[6] However, this included all types of immunodeficiency, both cellular and antibody, as well as minor antibody deficiencies (such as IgA deficiency and IgG subclass deficiency) which rarely need treatment. Differences in practice in relation to IgRT have been documented between European/ PID-focussed American immunologists, and general allergy and immunology

physicians in the US who deliver a substantial proportion of IgRT, and this could account for the high usage of IgRT in this survey. [7]

Common Variable Immunodeficiency (CVID) is the most common PID requiring IgRT, and in most series accounts for approximately half of all patients receiving IgRT. A recent Danish national survey confirmed the prevalence of CVID to be 1:26,000. [8]

A recent survey of patients receiving IgRT in Ireland demonstrated that there were 223 adults receiving IgRT under the care of immunology teams, 99 of whom were treated in the immunology centre, 32 in local hospitals and 92 were undertaking home therapy. [9] Updated figures (Sept 2019) indicate 286 adults, of whom 114 are on home therapy, 113 in immunology centres and 46 in local hospitals. Additionally, there are 52 children with antibody deficiency receiving IgRT (15 hospital based and 37 on home therapy). Hence there are currently a total of 338 patients in Ireland, who are included in the scope of this Model of Care.

1.4 Benefits of Treatment

Licensed immunoglobulin products have demonstrated reduced infection rates after immunoglobulin replacement therapy has been initiated when compared to pre-treatment rates. [1] In line with FDA guidance placebo controlled trials are not permitted given the importance of this therapy in antibody deficiency states. In many primary immunodeficiency disorders there is no alternative to immunoglobulin replacement therapy. [1]

Immunoglobulin replacement therapy has been conclusively shown to reduce the frequency of bacterial infection, reduce antibiotic usage, reduce fever and days off school / work and reduce hospital admission. [10,11,12] Treatment is targeted towards infection prevention, with individual patients requiring different doses and trough levels to stay infection free. Patients with immune deficiency complications such as bronchiectasis and gut protein loss may require higher doses of immunoglobulin to maintain individualised adequate trough / steady state IgG levels.[13]

The consequences of delayed diagnosis and inadequate treatment of antibody deficiency disorders are clear. Studies repeatedly demonstrate that diagnostic delay is common, typically 6-10 years from onset of symptoms.[14,15] Early diagnosis reduces pulmonary morbidity and prevents bronchiectasis, preventing long term negative cost implications.[16] Prompt diagnosis and treatment prior to the onset of complications is compatible with near normal life expectancy. However patients who develop complications have a significantly impaired survival.[17]

1.5 Objective

The aim of this document was to develop a model of care for IgRT in patients with PID, outlining recommendations for the standardised approach to the treatment and management of IgRT, and the conditions for hospital involvement.
This model of care is in line with the fundamental principles of Slaintecare. [18]

Patient is Paramount:

- Home therapy for suitable patients is the treatment of choice for PID and is associated with improved Quality of Life, and the opportunity to engage fully in education and employment
- Home therapy is associated with a lower rate of infections,

Timely Access:

- Moving patients out of hospital infusion rooms will allow patients awaiting infusion therapy access the care they are awaiting

Prevention & Public Health:

- By treating patients promptly, long term structural end-organ damage, particularly bronchiectasis can be avoided.
- When patients receive adequate therapy in a timely fashion, before the onset of complications, patient survival is virtually normal.

Free at the Point of Delivery:

- Current packages of treatment are free at the point of delivery.

Workforce:

- After a period of training patients become experts as self administration. Remote support is required, however this is less time consuming that administering infusions.
- Staff, in an area of severe skills shortages, are freed to assess and treat other patients.

Public Money and Interest:

- There is ample international literature to show that home therapy is the most cost-effective form of IgRT
- Freeing staff and hospital capacity to allow treatment of other patients will positively impact on waiting lists

Engagement:

- A major source of frustration for nurses and doctors in immunology is the lack of a process to allow us offer optimal therapy to our patients. Facilitating progression to home therapy for those waiting will improve staff engagement
- Patient engagement is significantly increased when patients become true partners in managing their chronic disease

Accountability:

- This initiative proposes delivery of optimal care with improved value, rather than second best care at higher costs.
- Governance of clinical care remains with the clinical immunology team

1.6 Need for Model of Care

Home therapy is associated with improved health-related quality of life, reduced costs.[19] and recent data suggests, reduced infection frequency.[20] During a recent review of IgRT practice in Ireland, marked geographical variation in access to home therapy was identified. Rates of home therapy provision among the 3 adult immunology centres were between 18% and 60%.[9]

The primary barrier to home therapy for suitable patients was availability of funding through the local health office. Other barriers identified were the lack of Immunology Clinical Nurse Specialists/Advanced Nurse Practitioners, as well as insufficient Consultant Immunologist numbers.

Immunoglobulin providers are approaching non-immunology specialists, offering to arrange training and supply of immunoglobulin to their patients at home. However it is recommended internationally and by this model of care that patients with PID should be seen by immunologists.[1] There is recognition that IgRT is more likely to be prescribed in line with guidelines by PID-focussed clinicians, compared to those where PID accounted for <10% of their practice. [7] To ensure optimal outcomes, patients need to be treated within

established governance arrangements with ongoing oversight, monitoring and support from an immunology/ haematology team, trained and experienced in the provision of home therapy. The need for care by PID specialist teams, together with the need for training for self-infusion by suitable patients at home, with regular follow-up to ensure on-going high standards was recognised in a recent International Principles of Care document, supported by the International Patient Organisation for Primary Immunodeficiency. [1]

In order to allow optimal therapy of patients with PID, it is essential to organise services such that there are no administrative barriers to accessing community supply of subcutaneous immunoglobulin. Ireland is the only country in Western Europe where there are still limitations in accessing subcutaneous immunoglobulin for suitable patients.[21] This forces immunologists to continue to treat patients with more expensive, hospital based infusions, with negative consequences for patients' quality of life.

Fig 1. Availability of subcutaneous immunoglobulin, a marker of home therapy availability across Europe. From Šedivá et al., 2014 on behalf of the European Immunoglobulin Map Group [21]

This Model of Care details:

- the indications for IgRT in PID,
- options for delivering IgRT, and
- requirements for safe delivery of Home Therapy

2. Immunoglobulin Replacement Therapy

Therapeutic immunoglobulin is a limited, relatively expensive resource and intermittent interruptions of supply occur because of occasional manufacturing difficulty or quality control failures. The UK Department of Health (DoH, England and Wales) introduced clinical guidance on the use of IgRT, which covers approved indications, recommended dosing and monitoring.[22] These guidelines are based on the strength of evidence, expert opinion and the availability of alternative therapies for medical conditions. Based on this classification, DoH approval for the use of IgRT is automatically granted (red), approved when supply is not compromised (blue), granted if alternate therapy unavailable or ineffective (grey) and not normally granted (black).

2.1. Indications for Treatment

Immunoglobulin replacement is indicated in patients with significant, symptomatic defects of antibody production or function, and for most patients IgRT is a lifelong requirement.[20,21]

Primary antibody deficiencies may arise due to a known or suspected genetic defect or in some patients with complex medical conditions which adversely affect the immune system either directly or through management of their underlying condition. [20,21]

These conditions include

- Common Variable Immune Deficiency (CVID)
- X-linked Agammaglobulinaemia (XLA)
- Germinal centre class-switch recombination defects
- Combined immune deficiencies including severe combined immune deficiency (SCID) and unclassifiable disorders
- Other primary antibody deficiencies including unclassifiable disorders
- Specific antibody deficiency (SPAD) with recurring or severe infections despite antibiotic prophylaxis
- Good's syndrome (combined immune deficiency with immunoglobulin deficiency)
- Some infants with prolonged physiological delay in native antibody production leading to significant infections (may not require lifelong therapy)
- Syndromic immunodeficiencies (e.g. 22q11 deletion syndrome, Trisomy 21, Jacobsen syndrome)

Of note all of these indications are “red” indications according to the UK immunoglobulin management guidelines, indicating that approval is automatic, for long-term treatment, as these patients have no other effective treatment option.[22]

2.2. Treatment Options

Currently licensed immunoglobulin products in Ireland can be divided into:

i) 5% and 10% intravenous products
ii) 10%, 16% and 20% subcutaneous products
iii) Hyaluronidase-facilitated subcutaneous products

These products have shown similar efficacy in terms of protection against infection [2, 24,25,26,27] however they are not identical with differences in properties such as IgA content, pH, additives (stabilisers), sodium content and osmolality due to different manufacturing methods. These differences could lead to differences in tolerability and adverse effects. Therefore, once a patient is established on a specific immunoglobulin product, this should not be changed to a different product in the absence of a clinical reason, without consultation with their clinician. [27]

Route of administration

IgRT can be administered intravenously, or subcutaneously. Subcutaneous therapy can be administered using infusions pumps, by rapid manual push, or by hyaluronidase facilitated large volume subcutaneous infusion. In Ireland, the majority of patients receive immunoglobulin either by the intravenous route, or by traditional, pump-delivered subcutaneous administration. However, facilitated subcutaneous immunoglobulin, either at home or in hospital, is increasingly used. Internationally, regular push subcutaneous administration of immunoglobulin is also being used. Some of the main differences between the different routes of administration, which impact on patient choice are summarised in Table 1.

	Intravenous	Facilitated Subcutaneous	Pump-delivered subcutaneous	Rapid-push subcutaneous
Frequency of infusion	Usually 3 weekly	Usually 3 weekly	Usually weekly	Usually alternate day
No. of needles	1	1	3-4 depending on dose	1-2
IV access required	Yes	No	No	No
Infusion Partner required	Yes	Yes	No	No
Equipment required	Drip stand	Drip stand	Pumps x 2	None
Treatment duration	3-4 hours	2-3 hours	1-2 hours	10-20 mins
Suitability for long term hospital therapy	Yes	Yes	No – frequent infusions	No – frequent infusions
Suitability for home therapy	Only for small no of patients	Excellent– with infusion partner	Excellent	Excellent

Table 1. Features of different types of IgRT which frequently impact decisions about long term management

Intravenous immunoglobulins became available in the 1970s, and allowed administration of adequate doses of immunoglobulin to patients with PID, such that serum immunoglobulin levels could be restored to within normal reference ranges. This was associated with improved outcomes and survival for patients, compared with the limited replacement which had been possible with previously available intramuscular preparations.[17] Intravenous immunoglobulin is typically administered every 3 weeks, and produces a high peak level of IgG, which falls rapidly over 1-2 days, following which the level falls slowly over 3 weeks. Adequacy of therapy is assessed by clinical response and monitoring of trough IgG level. Initiation of IgRT with IV infusions allows normalisation of IgG level much more rapidly than with subcutaneous immunoglobulin. Infusion side effects

are common and may be severe. Infusion reactions are often related to rate of infusion, and the presence of intercurrent infection. Early infusions typically take approximately 6 hours, however once the patient reaches maintenance infusions this is reduced to 3-4 hours. Unfortunately, some patients may continue to require slow infusions, due to frequent reactions. Infusions need to be deferred when bacterial infection is present, until the patient has had 48 hours of antibiotic therapy, which results in scheduling challenges.

Subcutaneous immunoglobulin forms a depot, which is more slowly released into the blood stream, maintaining much more consistent serum IgG levels. Subcutaneous immunoglobulin may be delivered with the assistance of pumps, or using a rapid push technique.

When **pump-delivered, subcutaneous** immunoglobulin is usually infused weekly. Patients are normally equipped with 2 pumps, and deliver immunoglobulin at 3-4 sites depending on dose. Typically, infusions take 60-90 minutes for an adult. Local irritation at the infusion sites is common, but rarely leads to discontinuation of therapy. With the exception of site reactions, side effects are less frequent than with IV administration, and are rarely severe. There is no need for cannulation, and although it is preferred to have an infusion partner, with certain precautions, stable patients may administer subcutaneous immunoglobulin without an infusion partner present.

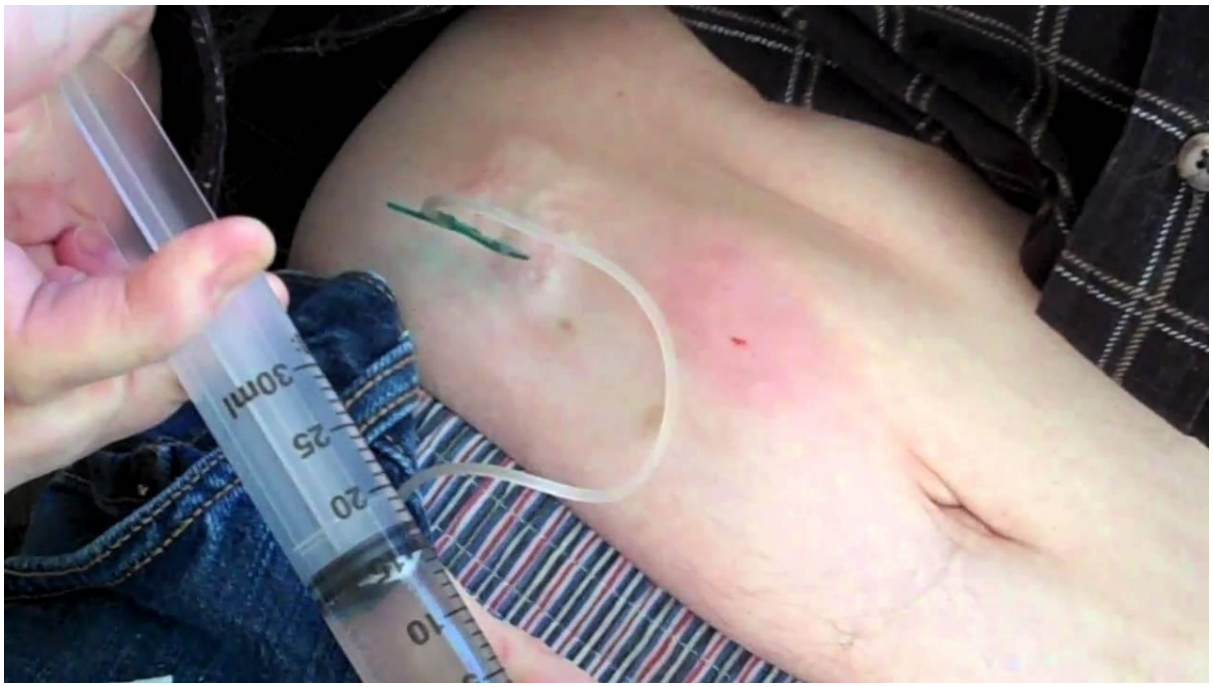

Fig 2. Appearance of site after rapid push subcutaneous immunoglobulin

Rapid push administration of subcutaneous immunoglobulin offers patients further choice. The immunoglobulin preparations are the same as those used for traditional, pump-delivered subcutaneous immunoglobulin. Up to 20 mLs of immunoglobulin is administered by slow subcutaneous injection, usually taking 10 minutes to administer. A single site is used, but treatment is given several days a week – often on alternative days. Other than site irritation side effects are rare. This method of administration has been adopted by patients who find it difficult to identify a consistent 1-2 hour slot each week to administer traditional subcutaneous immunoglobulin. Trough IgG levels and clinical response have been shown to be comparable to traditional subcutaneous administration. [28]

Site appearance is similar after pump delivered and rapid push immunoglobulin. The viscosity of immunoglobulin solutions require considerable manual strength to draw up, and in particular to administer by rapid push.

Facilitated subcutaneous immunoglobulin is a relatively new method of administration which has been licensed for less than 5 years. A subcutaneous injection of recombinant hyaluronidase is administered high in the abdomen, followed by an infusion of 10% immunoglobulin, manufactured to the same purity as intravenous preparations. Hyaluronidase breaks down hyaluronic acid in the subcutaneous fat, opening up space for administration of a large dose of immunoglobulin with a single needle. The infusion of immunoglobulin pushes the hyaluronidase through the subcutaneous tissue at the leading edge. A larger, less tense administration site develops. As the hyaluronic acid in the subcutaneous tissue turns over every 24-48 hours, the subcutaneous tissue is rapidly repaired.

Both intravenous and subcutaneous administration confer similar efficacy, each has its own advantages and disadvantages. The choice between these methods of administration is made based on several factors including clinical need, patient suitability, patient's tolerability of intravenous products, history of adverse effects, intravenous access and underlying medical conditions. [28]

Table 2: Advantages and disadvantages of intravenous vs subcutaneous route (29,30,31,32).

	Intravenous	Subcutaneous
Advantages	Less frequent infusions (usually every 3 weeks, but some patients may need more frequent infusions as per clinical outcome) Ability to reach a high serum IgG level more rapidly	Adverse effects infrequent, less than iv route Suitable for patients with difficult iv access Shorter infusion time
Disadvantages	May cause "wear off" effects in some patients with increased infection risk, fatigue towards the end of dosing cycle due to drop in IgG serum trough level Usually hospital based or infusion centre based (although home therapy is possible in some patients)	No "wear off" effects as steady-state IgG serum level is achieved with subcutaneous infusions More frequent infusions with multiple sites / infusion May not be suitable in patients with severe widespread skin disease Local site reactions are common

2.3. Hospital or Home Therapy

IgRT may be delivered in hospital or at home. Home therapy is associated with reduced healthcare costs, and improvements in multiple domains of health-related Quality of Life (see below). While the reduction in costs is primarily due to the reduction in nursing and administrative costs associated with regular hospital admission [33] a detailed French economic evaluation demonstrated that while theoretical cost-minimisation models showed a small cost reduction in favour of home based subcutaneous IgRT, that field data showed a 25% reduction in costs in stable patients because of lower doses required to give satisfactory trough levels.[34] Cost minimisation studies in France, [34] Canada,[35] Germany,[36] Switzerland,[37] and Japan[38] have all found in favour of home subcutaneous IgRT compared to hospital based IV IgRT. This has been confirmed by a metaanalysis of the available literature.[39] A pharmoeconomic evaluation in Japan showed a 55% reduction in productivity loss following a switch from hospital based infusion therapy to home-based

subcutaneous therapy.[38] A labour force analysis reported from Canada showed a reduction of one whole time equivalent nurse for every 37 patients moving from hospital based infusion to home therapy.[40] Of note this study considered the nursing time required to train, monitor and support patients in the home setting.

2.3.1 Hospital Care

When administered in a hospital setting IgRT is most commonly delivered by the intravenous (IV) route, as administration is only required every 3 weeks for most patients. Facilitated subcutaneous immunoglobulin is a suitable alternative for hospital based administration, as dosing interval is similar, and is a useful alternative for some groups of patients [40] including those with poor venous access. (Immunodeficiency is a relative contraindication to the insertion of permanent venous access devices due to the increased risk of infection). Additionally subcutaneous administration, (either traditional or facilitated subcutaneous) is occasionally required for patients who have repeated severe reactions to IV immunoglobulin, which cannot be prevented by careful treatment of infection and rate management, or those with protein losing enteropathies. [41]

The majority of patients in Ireland and the UK commence IgRT using IV immunoglobulin, as this achieves normal immunoglobulin levels more quickly. The incidence of side effects is highest in the first few infusions, and therefore close monitoring during administration in hospital increases safety.[27] This period of close contact with the immunology team facilitates optimisation of other aspects of the patients' management, allows ample time for the patient to ask questions and learn about their condition and management. Discussion about and assessment for suitability for home therapy commences in parallel. Where patients live long distances from the immunology centre, and are not suitable, or not happy to consider home therapy, arrangements will be made for them to infuse in a local hospital, while remaining under the governance of the immunology centre in terms of follow up, and recommendations for immunoglobulin dosage and rates of administration. Hospital based therapy with its higher overall costs, is typically reserved for those patients (eg. unsupported elderly with numerous comorbidities) in whom home treatment is impractical and unsustainable. [28]

2.3.2 Home Therapy

Patients receiving IgRT at home may use intravenous, facilitated subcutaneous, subcutaneous (pump-delivered), or rapid push subcutaneous immunoglobulin. Throughout Europe subcutaneous immunoglobulin therapy is the modality of choice for replacement therapy at home, due to the lower rate of adverse events, improved ease of administration, improved quality of life indices and reduced overall costs. [42,43]

Independent of the route of administration, well documented advantages of programmes for self-infusion at home include: [1]

- Adult patients report that they are less tired, can plan their lives and do not have to miss work to attend treatment sessions
- Parents report that home therapy keeps the child healthier due to regular treatment, enabling participation in school activities
- Participation in family/social and leisure activities for adults and playing with friends for children allow them to feel and act like others
- Parents themselves report less worry for the future of their child, fewer restrictions or sudden changes in plans in relation to family activities (eg holiday trips), less tension at home and more time for their own needs, and therefore a higher quality of life
- Improved flexibility for travel, both work and leisure, as patients can take their subcutaneous immunoglobulin with them.

2.3.3 Quality of Life

A 2004 quality of life study of both adults and children undergoing SCIG therapy in Europe and Brazil found that children who changed from hospital to home-based Ig therapy experienced improved school and social functioning, as well as there being a reduction in emotional distress of their parents. It also found that there were fewer limitations in the families' activities. The adults who changed from hospital to home-based therapy reported an improvement in social functioning, mental health, and general vitality. [44] Henderson (2003) reporting on a patient satisfaction survey of home Ig therapy undertaken at Papworth Hospital NHS Trust found that overall the patients benefited from home Ig therapy.[45] Patients reported being more relaxed, and that home therapy was more flexible and could be fitted in with their daily routine. Patients also reported that there was less disruption to the family. Positive aspects of home Ig therapy for immunodeficiencies , such as independence, flexibility, freedom and control, have been reported by patients in other reports. [44,46,47,48, 49,50,51] The further positive qualities of self-esteem and confidence,

has encouraged other inpatients to consider home Ig therapy as an option for their long-term management. [46,49,52] Negative qualities brought to the fore with home Ig therapy are few and far between, but Abrahamson, Sanderson and Bustnes (1996) mentioned the anxieties and worries of parents who had children undergoing home therapy, as they assumed greater responsibility for the medical care of their children.[46] Another benefit of home Ig therapy for antibody deficiencies is reduced cost - both to the hospital/health service as well as to the patient/family. The cost benefits to the hospital/health service include reductions on the time of health professionals, plus less use of physical services such as hospital beds. For patients and families, the cost benefits include not having to take time off from work and a reduction in fares to the hospital or health centre. [45,46,48,50,52] Home therapy is associated with a significant reduction in school absenteeism in children, and minimises the social and educational disruption associated with hospital based IgRT. This in turn reduces the costs of educational and other supports which the child will require.

A Beaumont hospital satisfaction survey of patients on the Immunoglobulin home therapy programme (2008) found that most were happy with the information received and the ongoing support from the Immunology team while all were happy with the training programme and preferred the home environment for infusions. A further Beaumont audit of the home therapy service in 2013 found that at the review appointments home therapy was having a positive impact on participating patients. Most patients expressed a desire to stay in the programme, comments compiled in the review process include;

"I feel better life is better"

"I have been able to get back to work"

"It allows flexibility in my life"

"I do not want to come to the hospital for life"

A further study conducted in St James Hospital assessed patients' knowledge and attitudes to home therapy, in this cohort patients were also extremely happy with home infusion and felt that it kept them well or very well and allowed them to participate in activities of daily living. Overall, 95% liked self-infusion quite a bit or very much with 70% finding them convenient [53]

3. Patient Perspectives

Patients attending Immunology services were invited to contribute to this document, by writing a piece of up to 500 words summarising the impact of diagnosis of their immunodeficiency and treatment, and for those who have been able to access home therapy, how this form of treatment had made a difference to them. An invitation was also extended to members of the Irish Primary Immunodeficiency Association to contribute.To preserve the confidentiality of those patients who kindly agreed to share their personal stories, names have been changed.

Mary (Age 32)

Throughout my childhood and teenage years I suffered with recurrent skin, ear, eye, chest, sinus and throat infections. I was exhausted all of the time. I was often in the GP surgery, took antibiotics up to five times a year and missed a lot of school. In 2002, when I was seventeen, I had a particularly long stay in hospital with fever, enlarged lymph nodes and granulomas. This stay in hospital culminated with the loss and removal of one of my organs, my spleen. I was lucky to get a diagnosis in 2003 of Common Variable Immunodeficiency Disease (CVID), thanks to the efforts of an excellent consultant immunologist. My immune system does not function as it should. My body does not produce enough serum immunoglobulins and antibodies, meaning that I am more susceptible to infections. This diagnosis changed my life. Home therapy has allowed me to be a fully productive member of society and lead a normal life.

Why do I need immunoglobulin?
Immunoglobulin replacement therapy is essential to replace the part of my immune system that is missing. Without this therapy, I would get recurring infections that could eventually lead to further scarring and a chronic debilitating condition, such as pulmonary lung disease. I would be very sick, very often. Both my quality of life and life expectancy would be greatly reduced if I did not have access to this treatment.

Why intravenous immunoglobulin at a hospital did not work for me.
Immunoglobulin replacement therapy that is self-administered at home is the only treatment that allows me to have a reasonable quality of life. I spent a year (2003-2004) travelling to Beaumont Hospital every three weeks for a nurse to administer immunoglobulin intravenously into my body. This infusion took up to six hours and could only take place during business hours. This was not the best treatment available for me, for the following reasons.

Firstly, my health was not as good on intravenous immunoglobulin. I suffered from peaks and troughs. Before my infusion was due, my levels would drop and I got tired

and sick. After my infusion my levels would soar, often causing reactions. I also have very small veins, meaning the trainee doctors struggled to put a line into a vein on my arm. On one particular occasion, this task took four trainee doctors a whole hour and left me bruised, in pain, crying and traumatised. In addition, forcing a person who has a very compromised immune system to come into a hospital on a regular basis for a considerable length of time introduces this person to unnecessary risk of picking up an infection e.g. flu, MRSA. I find this counterintuitive. I need to protect myself from viruses and bacteria. The thought of losing my home therapy fills me anxiety, fear and stress.

Secondly, intravenous immunoglobulin requires one day in a hospital every three weeks. This makes it difficult to hold a normal job and have a normal career. I would lose the job I currently have if I was forced to go to hospital for treatment every three weeks. Hospital schedules are inflexible and day wards do not open in the evenings or weekends. If I had to find a new four-day-a-week job, I would lose 20% of my salary, significantly reducing my disposable income. It would also reduce some social contributions and pension entitlements.

Future

I currently administer immunoglobulin subcutaneously at home every Saturday. Although this form of administration is better than intravenous treatment, it does impacts on my life. I do my infusion on a Saturday because I found doing the treatment when tired (e.g. in the evenings after work) increases the severity of reactions. The day after treatment, I can have fatigue and flu like symptoms. I call this my 'infusion hangover'. I need the Sunday off-work to recover. My consultant has said that I am suitable for a new medicine, which is a subcutaneous infusion that you only need every three weeks. I am hoping to switch as soon as possible. This new product would give me back most of my weekends and reduce the amount of days I struggle with fatigue, headaches and flu-like symptoms.

Summary

Being able to treat myself at home has meant that I can fit my treatment and medicine into life. Treatment does not take over my life. I have been able to hold down a good full-time job, earn enough money to ensure I eat well and take care of my health. I can control my environment and reduce the risk of picking up new infections. I can be a proactive patient and have learnt how to keep myself well. I have never been healthier than I am now on subcutaneous infusions. I take less sick days than my colleagues. Having access to a 3 weekly subcutaneous treatment would improve the quality of my life even more. Ultimately, self-administered immunoglobulin at home has allowed me be healthy, lead a normal life and contribute to society.

From the father of Keith (Age 19),

1. Why immunoglobulin replacement treatment is needed?

His diagnosis is DAVID Syndrome due to a Gene mutation in the Gene called NFKB2 – it's a rare condition combining anterior pituitary hormone deficiency and Common Variable Immunodeficiency (CVID). He has none or very little Immunoglobulins (Ig) IgG, IgM, IgA, IgD and IgE. Therefore he is unable to produce adequate amount of Ig, in other words, antibodies. He must have antibody replacement to be able to live and to have a good life quality.

2. How subcutaneous treatment at home makes a difference?

He used to receive the intravenous Ig replacement, however, not only had many side effects, he was very sick for the first 48 hours, nauseas, headaches and skin eczemas. Also he was unable to maintain the Ig level in the blood from one session to another. He was only able to maintain levels for the 10 days, treatment was every 21 days. After the first 10 days he was very venerable always sick, pneumonias, virus, meningitis, etc. and in need of going to A&E, hospitalising, etc. He developed bronchiectasis on his right lung, doctors indicated that the only way for him to have a better quality of life he should be on weekly subcutaneous therapy. He has been in this home therapy for the last 5 years with excellent results. He has been able to go to school and enjoy today a normal teenager life, with very few episodes of illness. Therefore the subcutaneous treatment at home gave him the opportunity to be able to live just like any other boy. Which is his right as a human being.

Una (Age 20)

In 2016, I was rushed to hospital. I was physically unable to breath. I spent the night in isolation in A&E and diagnosed with severe pneumonia that had caused my lung to collapse. I now know that pneumonia is on the list of infections CVID patients are at high risk of contracting. Since 2016 I've been on 8+ antibiotics each course lasting 10 consecutive days along with 3 hospital admissions.

Common variable immune deficiency is complicated. Just before my treatment date symptoms begin to raise their head. The only possible way I can describe these symptoms to somebody that is lucky enough to never have to experience them is that you are so exhausted you feel like you are dreaming, like you're not really present. And of course, the few days before treatment is when your immune system is at its weakest and nine time out of ten you have the joy of being plagued with infections.

Visiting the hospital every three weeks for treatment is, honestly, painful. Due to your antibody levels plummeting your body is working harder to continue to function 'normally', leaving you exhausted and sick. On these days, I have to get up early, even though my body refuses to cooperate, and travel to Dublin to sit in a day ward for the majority of the day.

Since my diagnoses I have been receiving subcutaneous immunoglobulin. I was given the option of intravenous immunoglobulin but since I am young and in college and living in Meath the consultant recommended subcut so I would be able to treat CVID at home, working treatment around my schedule instead of me having to work around the hospitals day wards hours.

Subcutaneous immunoglobulin treatment would usually take 2 hours maximum but since I have to travel to Dublin for treatment it takes the entire day because I am not the only patient in the room. Patients with colds, flus and other infections take priority due to them needing consultations before starting their treatment meaning other patients with comprised immune systems are not only being exposed to additional unnecessary infections but are having to dedicate a full day to the hospital.

I am currently in my final year in university studying Irish and media studies and once I graduate I would like to start my career as a translator. At the minute, it is literally impossible to get a job because I have to attend the hospital every 3 weeks for treatment. If I had home therapy the number of infections I get would lower, I could attend all my lectures, I wouldn't have to decline invitations to social events with friends. I would be able to live like any other 20-year-old without the fear and anxiety of getting sick hanging over my head.

Susan Age 19 ,

Diagnosis: APS type 1

When I first started getting immunoglobulin infusions it was at the hospital. Every three weeks I had to spend the day as an inpatient getting this infusion. I was very grateful to be getting the immunoglobulin because I felt so much stronger and wasn't in hospital as much with sickness and infections. However, each infusion day left me feeling so tired and weary and I came to dread the days I had to go in especially when I was feeling so well.

When I got changed to home infusions weekly I soon realised how much more of my life I was getting back. Instead of a whole day in hospital and feeling tired mostly from having to travel an hour to get there and spend the day in hospital getting signed in, checked by the doctor and nurses, waiting for a bed, getting hooked up, waiting for the treatment to finish, travelling and hour home again I could now do my own weekly infusion at home and be finished within an hour and a half.

Getting my immunoglobulin infusions at home has been such a benefit to me in so many ways. For one, I do not have to take a whole day out of my life to spend it in a hospital. I can decide when I do the infusion so I can just do it in the evening where it is not disrupting my daily activities. Another benefit that home infusions has had on my life is that due to me not being in hospital as much I pick up less infections and viruses.

I believe that home infusions are the best idea for patients who are immunocompromised as it means they are not in hospital as much, and for all their benefits and how much we need them hospitals are full of infections and viruses and not the best places to be when you are trying to stay healthy.

James, 22yrs old

I am and have been a patient of St. James's Hospital for the last few years and before that I was attending Tallaght Children's Hospital since I was about 3 years old. I attend St. James's immunology department as I suffer from a condition called x linked agammaglobulinemia (XLA). I am forced to attend the hospital every three weeks in order to get this treatment otherwise my immune system wouldn't be able to function properly leading to severe infections and pneumonia (as I've suffered from more than once growing up).

I believe I have suffered an enormous loss by being denied funding for home therapy treatment. I have been restricted my whole life from things such as playing sports, going on

holidays as they would have to tie in with my treatment as one year I overstepped the limit and ended up in a hospital on a drip with a severe infection. I have had to miss out on school trips, sports days and the list goes on. Throughout school primary, secondary and now in college I've to miss a day every three weeks because of this treatment. It's conflicting with study and assignments as well as work and my own personal life.

I feel I'm at a huge disadvantage to others not in my situation and one particular topic I would like to highlight is my Erasmus year. This year was my designated year away from college which I slaved to obtain a place in a college in America. I believed home treatment wasn't too difficult for me to obtain as others in different constituencies had received it in the past without any complications. Not in my case though. From my understanding of the refusal of my application was frankly down to discrimination, because, of where I live. I was told there was no funding for me simply because of where I lived. This application was re submitted with documents and letters from a wide variety of people explaining and outlining how beneficial and critical for my life it was for me to get this funding to get home and treat myself there. Again, this was denied and all the work I had done and money invested in securing a position in the college in America had been completely and utterly wasted.

I had thought the older I got the more manageable my condition would be due to advancements in healthcare etc. but that's far from the case. I almost wish I was a child again attending Tallaght Hospital not having to worry planning a holiday in the summer or seeing all my friends jet off on J1 visas and touring Asia and most heart-breaking seeing your classmates living in America where I thought I would be right now not writing an email complaining about how unjust our health system is. Another example which to be quite honest had me deeply upset for a long time and only recently got over it was that after hearing I couldn't travel for my Erasmus later that summer my sister decided to do a graduate visa in America and packed her bags and just left which left me feeling down and upset and angry that I was at such a huge disadvantage.

It is my understanding from my consultant that everyone is entitled to the same treatment under some Health regulation, I wouldn't know off hand exactly what is it but to simplify everyone should be able to receive the same treatment not only in Ireland but across Europe. However, this isn't even implicated within Ireland and two bordering counties. I just can't comprehend why I do not fit the criteria to receive this treatment

4. Organisation of Home Therapy Services for Immunoglobulin Replacement

Following demonstration of safety and efficacy during the 1980s, home therapy programmes where the patient, parent or partner were trained to administer intravenous or subcutaneous infusions at home were introduced. [33, 48, 54,55,56] Home Immunoglobulin therapy is now common practice and is widely practised throughout the United Kingdom (UK), Scandinavia and Ireland where nursing structures are in place to manage the programmes.

4.1. Governance

The delivery of hospital based immunoglobulin replacement treatments and the facilitation of home therapy programs should be carried out in centres with expert medical and nursing teams. [1]

Home therapy programmes should be staffed by consultant immunologists and clinical nurse specialists/advanced nurse practitioners in immunology, with appropriate NCHD and administrative support.

PID are complex diseases with complex complications, and while IgRT is a vital component of therapy, holistic care should be organised in an integrated patient-friendly way. The immunology team require access to specialist respiratory medicine, gastroenterology and infectious disease support on a regular basis, as well as access to physiotherapy, clinical nutrition and psychological services.

Overall governance for IgRT remains with the Clinical Immunology team, whether IgRT is delivered in the immunology centre, local hospital or at home.

4.2. Patient Choice

It is essential that home therapy is a positive choice for the patient. No patient should be forced to have home therapy because of lower costs, or pressure on capacity in an infusion room. In the event that a patient enrolled on a home therapy programme opts to return to the hospital setting either on a short or longterm basis, this must be accommodated.

In an international survey of 300 respondents (patients and caregivers) involved in home therapy programmes in 21 countries, all respondents expressed a desire to reduce infusion frequency, the ability to administer these at home, self-administration, shorter duration of administration, and fewer needle sticks. [57] The results of this survey highlight the importance of providing access to different treatment options and modes of administration to ensure individual patient needs are best met.

4.3. Assessment of Suitability

Following the decision to commence Immunoglobulin treatment each patient is assessed individually to determine a suitable treatment regime. Home therapy is a treatment option which would be considered in the majority of independent persons.

The decision to enrol a patient onto the home therapy programme should be a joint decision between the patient, parents (in the case of children), their infusion partner (if required) and the multi-disciplinary team (Clinical nurse specialist and the Consultant).

Adequate time should be allowed for the decision on which type of treatment is most suitable for the patient. Home therapy is not suitable for every patient and a thorough discussion should take place before any final decision on treatment is made. Patients should be given ample time to assess the benefits to each type of treatment available to them. Information should be given in writing and patients should be given ample opportunity to ask questions about home therapy and time given to consider the choice with family members involved. Information given should be age appropriate and directed at the patients/parents level of understanding and literacy skills.

A patient should not feel obliged to take part in the home therapy programme; this should be discussed whenever home therapy options are discussed with the patient.

The criteria upon which enrolment to the home therapy programme are :

- The Patient must have the dexterity to perform the tasks involved , in certain circumstances aids can be provided but the wish to commence treatment should lie with the patient
- The patient must have the required knowledge regarding their illness and treatments

- Demonstrate motivation and willingness to comply with the home therapy programme and all its implications and responsibilities and to sign a consent to this effect.
- Demonstrate compliance with medications and have a good record with attendance for hospital infusions and appointments.
- Have a supportive infusion partner who will be present for all home infusions and can attend for training;
    - On one occasion if the partner is solely in a supportive role
    - For all training sessions if in an active role
    - In the case of children, it is essential to have the commitment of both parents, where applicable.
- Have access to a landline telephone or reception on a mobile phone at the place of infusion
- Good venous access if having intravenous immunoglobulin.
- Have received immunoglobulin therapy in a hospital setting
- Written confirmation of funding from the HSE.
- Have successfully completed a competency assessment
- Confirm that any house pets are not to be in the room during set-up of infusions and preferably do not generally have access to this room.
- Agree to return to hospital based therapy if requested

4.4. Training

The Immunology nurse is responsible for the education, training and preparation of patients to participate in therapeutic self-care. Home immunoglobulin therapy is safe where there are guidelines for administration and where good education of patients and their families occurs prior to commencing home Ig therapy. [44,57, 58,59,60,61,62]

Elements/stages of home therapy training include;

- Introduction to home therapy, safety and procedure for infusing Immunoglobulin at home
- Record keeping, drug preparation, priming the line
- Drug administration, calculation of rates, pump management
- Insertion and removal of needle device, disposal of equipment
- Blood sampling (where appropriate)
- The prevention and management of adverse reactions

- Obtaining supplies, patient contact with the Immunology Department, Delivery company
- Assessment of competence

With regard to the paediatric population, training provided will be appropriate to the child's developmental stage. Parents or guardians will be provided with adequate support to allow for involvement in self-care when deemed appropriate.

There are a number of approaches to patient training. Many patients are exposed to the stages of home therapy by a gradual process each time they attend for their infusion. Patients will then either attend a fixed training programme in the Immunology centre, local hospital or the home setting. The patient and family member gradually become more confident with active participation and support with nurses who are specialised in home Immunoglobulin therapy.

4.5. Assessment of Competence

Assessment of competence will be an integral part of the training programme. At each infusion stage of proficiency will be assessed and documented.

In the paediatric population, assessment of patients and caregivers will be in line with the child's developmental stage and provision for self-care made with regard to the child's willingness to learn and ability to perform tasks. Transition from child to adult care will be discussed as part of a multidisciplinary approach and the child's learning need discussed with parents or guardians.

Patients will be allowed sufficient time to gain adequate knowledge and will be supported by the immunology nurse at each stage of training, a competency based model may be used to determine the stage of proficiency ie: Benner model, assessing from novice to expert or proficient. Competence should be documented and any outstanding learning needs addressed prior to the patient self-infusing at home

Assessment of competence should be discussed at review appointments and supervised infusions performed if indicated.

Competence should be assessed if, following review there may be medical, personal or psychological changes in the patient's condition.

Areas to be assessed to ensure competence include but are not limited to

- Rationale of the home therapy programme
- Indications and contraindications for Immunoglobulin use

- Ability to set up materials and equipment to perform infusion
- Adequate hand washing and awareness of infection prevention measures and their importance
- Checking of the Vial and dose , including recording of product infused and importance of documentation of such product
- Drawing up the infusion , where applicable
- Choice of site of infusion and rational for same
- Ability to use any ancillaries and equipment required to perform infusion, including trouble shooting
- Ability to trouble shoot any issues that may arise during the infusion
- Ability to discontinue the infusion and safely dispose of all used equipment with particular emphasis on safe management of sharps.

4.6. Support

On going support forms an integral part of the home therapy programme. Support at home is provided in a number of ways.

All patients and their carers have access to the immunology nursing team for queries and issues arising from their treatment. Hospital based nurses provide assistance and are a liaison with the medical and multidisciplinary members of the service. Adequate contact details should be available to the patient to ensure that they feel supported in the role of self administration.

Homecare nurses should be available to provide support for any issues that may arise that necessitate a home visit. Home visits must be provided by nursing members who work as part of the immunology service, either by means of a service level agreement or other agreement deemed appropriate by the supervising health care institution to which the Immunology centre belongs.

4.7. Review

Home therapy should be reviewed at six monthly intervals or sooner if clinical condition or home circumstances warrant such. Review intervals will be decided in conjunction with all members of the clinical team.

Review should consist of assessing competence and efficacy of treatment. Where issues are identified a full assessment of patient’s needs should be performed

During the patient's life cycle psychological, social and clinical needs may instigate the need for a change in treatment

Issues to be considered include

- Immunoglobulin product
- Method of administration
- Place of administration

Changes to patients' product, method of administration, route of administration or place of administration should be made in conjunction with all members of the clinical team.

5. Recommendations

5.1 When the decision is taken that IgRT is required, this should be started within 4 weeks, and no later than 8 weeks

Rationale:

The decision to prescribe IgRT may be taken rapidly in those with evidence of severe antibody deficiency, or after extensive work-up, often after failure to respond to prophylactic antibiotics. However prescribing IgRT means that no other treatment is available for the patient. The risk of developing bronchiectasis or sepsis increases the longer the patient remains without adequate therapy. Developing complications prior to diagnosis is associated with impaired long term survival.

Audit standards:

90% of patients commencing IgRT within 4 weeks of the decision to initiate therapy

100% of patients commencing IgRT within 4 weeks of the decision to initiate therapy

5.2 All patients who are suitable for home therapy should be offered training, and training should commence within 8 weeks of confirmation of funding

Rationale:

It is internationally recognised that home therapy is the optimal way to deliver IgRT, in patients who are suitable. Home therapy is associated with lower overall healthcare costs, and improved quality of life for patients. Home therapy also fosters a partnership approach to management with positive impact in other areas of care. Home therapy should be discussed with all suitable patients, no later than 6 months after commencing IgRT. Where a patient is not considered suitable for home therapy (eg personal or household substance abuse, severe co-morbidity, frequent infusion reactions), the reason for not offering home therapy should be documented. The outcome of home therapy discussion should be documented. Patients should be aware that even if not suitable at that time, home therapy may be an option in the future.

Once a patient has decided that they wish to undertake home therapy and funding confirmation has been obtained, training should be offered without delay. While it is often time-effective to train 2 patients together, and many patients find the resultant mutual support beneficial, no patient should be delayed more than 8 weeks prior to beginning training, given the negative impact of on-going hospital therapy.

Audit standards:

Home therapy discussed within 6 months of commencing IgRT in 100% of potentially suitable candidates.

95% of patients commence training within 8 weeks of the decision to pursue home therapy

5.3 Patients should have the right to choose the route of administration which is least burdensome, from medically appropriate options.

Rationale:

The immunology team should evaluate which route of home therapy is potentially appropriate for each patient. This should be followed by a discussion of the pros and cons of each potentially suitable method of administration, with the patient and potential infusion partner. The patient should have ample time to consider the options, before deciding on the choice of initial route of administration. The patient should be aware of the potential to change product or route of administration in the future, should the need arise.

Audit Standard:

Documentation of discussion of options in 100% of patients prior to applying for funding.

5.4 Barriers to home therapy should be addressed

Rationale:

Currently the biggest barrier to home therapy is obtaining funding approval. Even when funding is granted, time to approval varies between 4 weeks and several months, with different forms and supporting documents requested in different areas. Development of a consistent process for initiating home therapy should be put in place.

An additional barrier to home therapy is staffing in immunology units. While this has improved recently, in the event that an immunology unit cannot support training, arrangements should be put in place to obtain external support to assist with training, or referral to another centre for training should be arranged.

Geographical variation in access to Specialist Immunology Services can impact adversely on patients treated in such areas accessing all available therapy options and supports including home therapy. Development of adequately staffed specialist services to ensure equitable, national provision of access to home immunoglobulin therapy and expert support is required.

Audit standard:

100% of patients who are medically suitable and keen to participate in a programme should have arrangements put in place, regardless of geographical location.

All non-conformances should be monitored, and Quality Improvement initiatives put in place to prevent recurrence.

5.5 The governance of patients undertaking IgRT at home remains with their immunology home therapy centre. Appropriate support and review processes must be in place.

Rationale:

Antibody deficiencies are serious illnesses, and while IgRT is essential, other aspects of care need to be optimised. Patients need to be monitored for complications of their disease and therapy. Regular blood tests must be monitored to assess adequacy of therapy, and monitor

for complications. Home therapy technique and haemovigilance compliance must also be monitored.

Audit Standards:

100% of patients on home therapy must have a nominated home therapy centre, and details of how to contact home therapy personnel as required.

100% of patients should must monitoring of immunoglobulins, FBC and LFTs as a minimum. Failure to attend for blood tests may lead to discontinuation of home therapy.

100% of patients on home therapy must have scheduled out-patient appointments.

Acknowledgements

We are grateful to Dr J. D. Edgar, UK Primary Immunodeficiency Network for provision of current UK & Northern Ireland IgRT Data.

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Appendix 1

Model of Care Development Group

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Title of Guidance Development Group:	Members of the Irish Primary Immunodeficiency Association
Approved by:	Clinical Forum of the HSE Chief Clinical Officer
Unique Identifier Number:	CDI111/2019
Version Number:	1
Publication Date:	November 2021
Date for Revision:	November 2024
Electronic Location:	HSE NCPP webpage

Version	Date Approved	List Section Numbers Changed	Author