Pwyllgor Gwasanaethau Iechyd Arbenigol Cymru (PGIAC)
Welsh Health Specialised Services Committee (WHSSC)

# Specialised Services Service Specification: Specialised Immunology

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# 1. Aim

## 1.1 Introduction

The document has been developed as the service specification for the planning of specialised immunology for patients resident in Wales.

The purpose of this document is to:

- detail the specification for the specialised immunology services for patients who are resident in Wales;
- and identify which organisations are able to provide specialised immunology service for Welsh patients.

The approach to developing this specification has been grounded in the Public Sector Equality Duty principles of transparency, engagement, evidence and leadership to ensure that it impacts in a fair and positive way. An Equality Impact Assessment (EqIA) has been undertaken,and a separate document has been produced outlining the findings of the assessment.

WHSSC is committed to the planning and commissioning of services that are equitable, accessible and responsive to individual needs.

This document covers people who are affected by primary immunodeficiency disorders (PID) in Wales. Primary denotes the mainly genetic nature of the defects, differentiating them from secondary or acquired immunological disorders caused by malnutrition, infection (e.g., human immunodeficiency virus [HIV] infection), chemotherapy or other external agents. As stated this document focuses on primary immunodeficiency disorders (PID) and excludes secondary disorders and the use of IVIg to treat neurological conditions.

## 1.2 Relationship with other Policy and Service Specifications

This document should be read in conjunction with the following documents:

- WHSSC Specialised Service Policy for Blood and Marrow Transplantation

- The All Wales Policy: Making Decisions on Individual Patient Funding Request (IPFR) Policy. The IPFR Policy is available online via the Welsh Health Specialised Services Committee website: http://www.wales.nhs.uk/sites3/page.cfm?orgid=898&pid=59092

The Strategy currently being developed in Wales for rare diseases is also highly relevant for this service.

# 2. Service Delivery

## 2.1 Service Model

The service shall provide hospital-based outpatient and day-care with access to in-patient facilities. This will comprise:

- Regular outpatient clinics for assessment and follow-up.
- Adequate clinical space in relation to the number of patients being treated.
- Adequate space for patients receiving infusion or training.
- A safe working environment for staff.
- Access to an appropriately staffed designated day case unit that can provide immunoglobulin and biological infusions and training for home therapy both intravenous, subcutaneous and facilitated subcutaneous.

The service shall have access to support from other clinical specialties for complications of Primary Immunodeficiency including:

- Ear, Nose and Throat Medicine
- Respiratory Medicine
- Gastroenterology
- Infectious Diseases
- Haematology Transplant
- Haematology/ Oncology
- Psychiatry
- Paediatrics

- Clinical Genetics
- Rheumatology

The service shall deliver a diagnostic package comprised of routine and complex tests for the investigation for suspected immunodeficiency, including initial consultation and follow-up in a dedicated immunodeficiency clinic, specialised immunopathological tests in an accredited laboratory, test immunisations, specialised genetic and radiology studies: Specifically this will require:

- Accredited diagnostic services for the management of primary immunodeficiencies.
- Radiology and genetics +/- tissue typing.
- Specialised Immunology Laboratory services with CPA accreditation or equivalent.
- Access to diagnostics for rare and emerging diseases through European/USA laboratories.

The service shall have appropriate pharmacy and/or facilities including:

- Appropriate storage and dispensing facilities for drugs and immunoglobulin products.
- Pharmacy storage facilities for non blood product immunological therapies and documentation of dispensing to individual patients to allow reliable traceability.
- Blood bank or pharmacy storage facilities for blood product immunological therapies and to allow reliable traceability documentation of dispensing to individual patients.
- At the University Hospital of Wales Immunoglobulin and C1 esterase inhibitor are dispensed by blood bank rather than pharmacy and will need to meet MHRA requirement. All products are dispensed by Pharmacy at Manchester Royal Infirmary.

Adult Specialist clinical immunology services shall be provided by a multi-disciplinary team that includes:

- At least two Consultant Clinical Immunologists or single handed practice within a managed network with experience in management of patients with PID and who maintain up-to-date CPD in their area of practice
- Senior Specialist Nurses with immunology experience andtraining to provide nursing care, training and run the home treatment service. Senior Specialist Nurses should maintain CPDin their area of practice.
- There will be an iterative annual joint review of staffing levels by WHSSC and service providers to ensure appropriate caseload, with ability to increase staffing level in line with growth in patient numbers.

Transition arrangements and preferences should be discussed with the child and their family at least 12 months in advance, and in line with NHS guidance. Shared protocols between child and adult services should be established, defining the roles and responsibilities of each member of the teams.

In Wales there is a standard set-out within the National Service Framework for Children, Young People and Maternity Services http://www.wales.nhs.uk/sites3/Documents/441/EnglishNSF_amended_final.pdf specifically for the transition of children into adult services. The current UK guidelines for best practice in providing transitional services for young people as they move from children to adult services are under review by the Department of Health. A generic transition document is being developed.

*Manchester Royal Infirmary* has a Policy to support patients through the transitional period. Transition is audited at Manchester Royal Infirmary based on national standards. Manchester is in the process of revising and improving their policies.

*The University Hospital of Wales* work well as both the adult and children services work in parallel. When a patient reaches the point of transition the service works closely with the patient and the patient's family to develop a plan for transition.

*Liverpool & Broadgreen NHS Trust* maintain strong links with colleagues at Alder Hey Children's NHS Foundation Trust and work closely to ensure as smooth a transition process as possible. This includes development of joint clinics and introducing those patients coming up to transition to the adult service at the right time. We are currently reviewing our transition service in liaison with those patients who have gone through the process to improve our working practice in the future.

In addition to those services listed within the Service Model the provider shall also maintain links with the following services:

- Secondary care
- Community Paediatrics
- Genetics
- Play therapy
- Psychology
- Psychiatry

- Paediatric Rheumatology
- Cleft Lip & Palette
- Depending on the nature of the immune disease, services are involved in shared care in relation to general medical needs, delivery of antibiotics and, other medical thereapies (small number of patients receive Ig therapy at peripheral hospitals) with:
    - Primary care
    - Care plans/clinic letters of PID patients are shared with primary care.
    - Antibiotic guidelines are individualised and advice and treatment is shared with general practitioners via clinic letters.
    - Clinic letters are sent to GPs and other specialties involved in a patient's care.
    - Private sector and third sector links

The service shall maintain a strong liaison with the Primary Immune Deficiency Patients Group for Wales (IDPGW) and Primary Immunodeficiency UK (PID UK), and liaise with other patient groups as appropriate including the Chronic Granulomatous Disorder Society, Hereditary Angioedema UK, Genetic Disorders UK, Genetic Alliance, Wiscott Aldrich Society, Max Appeal (Di George Society), UK Primary Immunodeficiency Patient Society, etc.) to provide further community support and continuity of care.

## 2.2 Interdependencies with Other Services

The service shall have access to related services required for the optimal care of PID patients.

Clinical immunologists must liaise closely with colleagues in a range of specialties, including respiratory medicine, ENT surgery, dermatology, haematology, oncology, infectious diseases, gastroenterology, general surgery, genetics, ophthalmology, rheumatologists and behavioural medicine.

The service shall deliver close input from physiotherapy via the non CF bronchiectasis clinic - essential for the management of the respiratory complications associated with PID.

The service shall ensure access to social workers, psychologists and dieticians for selected patients as required for the holistic care required for PID patients.

For centres without paediatric immunologists, the service shall triage and identify patients requiring referral to highly specialised national services for immunodeficiency at Great Ormond Street and Newcastle.

The service shall deliver access to close support from an accredited diagnostic immunology laboratory providing a range of routine and specialist assays.

While most services do not have dedicated in-patient beds - access is available and admission pathways for PID patients should beestablished with individualised care plans where necessary.

## 2.3 Specialised Immunology Services for Welsh Patients

The following providers are commissioned through WHSSC for Welsh Patients:

- University Hospital of Wales, Cardiff & Vale University Health Board – South Wales
- Central Manchester Foundation Trust (adults & children)– North Wales
- Alder Hey Children's Hospital – North Wales

These providers provide the following services:

- Diagnosis, assessment and treatment of patients with suspected immunodeficiency, and for a proportion of patients to exclude the possibility of PID
- Hospital infusion clinics and specialist drug treatments
- Home training and monitoring for immunoglobulin replacement therapy
- Training, advice and links to infusion units in other hospitals in Wales where patients require infusions.
- Diagnosis and management of patients with hereditary and acquired angioedema, including home therapy training
- Specialist Clinics at University Hospital of Wales
    - MDT Clinic for Di George (22q11)
    - Paediatric BMT
    - Paediatric Fever Syndrome
- Specialist Clinics at Manchester Royal Infirmary and Royal Manchester Children's Hospital
    - Adult BMT Services for PID

- Paediatric Immunology service for PID including transplantation for non-SCID PID

Whilst the service is primarily delivered by University Hospital of Wales & Central Manchester Foundation Trust, there is also Welsh activity reported at Salford Royal and Alder Hey. Liverpool & Broadgreen also accept referrals from North Wales; however this is funded through contractual arrangements between Betsi Cadwaladr University Health Board & Liverpool.

In North Wales Children with severe PID are referred to the Royal Manchester Children's Hospital. As previously indicated there is also a flow of referrals into Alder Hey. This service is provided on an outreach basis and Alder Hey's acceptance would depend on the severity of the disorder. Adult referrals in North Wales are made primarily to Manchester Royal Infirmary & Liverpool & Broadgreen.

In South Wales Children are treated at the University Hospital of Wales, Cardiff & Vale University Health Board. The children's service is run in parallel with the adult service on the same site.

## 2.4 Highly Specialised Immunology Services for Welsh Patients

The following providers are designated to provide highly specialised immunology services:

| **Provider** | **Service** |
|---|---|
| Great Ormond Street Hospital NHS Foundation Trust | Paediatric Severe Combined Immunodeficiency and Related Disorders (SCID)<br>BMT<br>Gene therapy<br>Autoimmune Gut Diseases (AGD)<br>Juvenile Idiopathic Arthritis(JIA) |
| Newcastle upon Tyne Hospitals NHS Foundation Trust | Paediatric Severe Combined Immunodeficiency and Related Disorders (SCID)<br>BMT<br>Autoimmune Gut Diseases (AGD)<br>Juvenile Idiopathic Arthritis(JIA)<br>Specialist lab testing |
| Royal Free Hampstead NHS Trust | Cryopyrin Associated Periodic fever Syndromes |

| | |
|---|---|
| | BMT<br>Specialist lab testing |
| University Hospital Bristol NHS Foundation Trust | Barth Syndrome<br>Transplants for Paediatrics |

## 2.5 Shared Care

Arrangements for shared care should be individualised for each patient based on the need for, and availability of, local services through the local health board services within both primary and secondary care.

The communication and engagement between the Specialist provider and the local providers is significant to ensure that patients are managed appropriately and receive their infusions close to home when appropriate and as safely and effectively as possible.

## 2.6 Self Care

The service shall provide patient self-care as an option in their management based on the patient's wishes, abilities and circumstances, to include:

- Provision of information about when to seek advice from the PID centre about obtaining or taking antibiotics to training for the administration of blood products at home.
- Competency testing (for example after home therapy training).
- Provision of home therapy (a flexible approach to treatment) as a package of care on a named patient basis including nursing supervision, C1 inhibitor or immunoglobulin therapy (intravenous or subcutaneous), infusion sets, pumps for subcutaneous delivery, deliveries of consumables to patients' homes, regular outpatient consultations and monitoring of antibody levels, blood counts and liver function tests.
- Assisted homecare should be considered for patients unable to self-infuse, under the care of appropriately trained nurses.
- The provider shall ensure that all home care programmes are registered and applying for accreditation through UK Primary Immunodeficiency Network (UK PIN).
- Patients with confirmed PID requiring regular immunoglobulin replacement therapy will be provided with a management package comprising:
    - Day case attendance every 1-3 weeks, nursing supervision, drugs, intravenous (IVIG) or subcutaneous

(SCIG) immunoglobulin, pumps for SCIG, monitoring by specialised immunopathological tests, radiological imaging, lung function tests, biochemical tests, medical and nursing follow-up, monitoring for efficacy and adverse effects and control of this expensive/scarce product.

- Acute and long-term management for patients who require C1 esterase inhibitor (or other high cost parenteral drugs) for treatment or prophylaxis (e.g. surgical, dental or investigational procedures) including managing those patients on home therapy.
- Management of those immunological disorders requiring other/new treatments (e.g. monoclonal antibodies or cytokines) on a named patient basis, where there is asuitable evidence base or need. This includes day caseattendance, nursing supervision, the drug, pumps for subcutaneous or intravenous use, monitoring by biochemical tests, specialised immunopathological tests and medical follow-up.

## 2.7 Hospital Administered Infusions in DGHs

Where it is clinically appropriate, the service nursing team will liaise with local hospitals to ensure that patients are able to receive their infusions closer to home. Patients who are treated closer to home may require admission to the specialist centre at some-point due to the complex nature of their condition.

## 2.8 Care Pathway

Referrals can be made from both primary and secondary care as follows:

- Due to the complex nature of PIDs, tertiary referrals into the immunology services come from Tier 2 (general physicians) or other Tier 3 tertiary or specialist physicians (particularly respiratory, ENT, gastroenterology and haematology).
- Primary Care Physicians (Tier 1) may also refer patients directly to the service, though these cases will require screening by the service to ensure the referral requires specialist input. A care pathway with referral guidance should be developed.

A referral pathway has been developed and is attached as appendix A.

## 3. Scope

### 3.1 Aims and Objectives of the Service

The provider shall ensure that Specialist Immunology centres will provide:

- A high quality, accessible and sustainable service that meets the needs of the population within its catchment area and reflects effective resource use and incorporates the views of patients.
- Equity of access to best practice standards, based on current guidelines for diagnosis and management for patients with PID and related complications.
- Excellent, holistic, multidisciplinary care for patients with immunodeficiency, complex autoimmunity and auto-inflammatory syndromes according to best practice guidelines defined by UKPIN, (European Society for Immunodeficiencies) ESID and other authoritative bodies.
- Integrated care with primary, secondary and other care providers and ensure close links and collaboration with other expert centres at national and international levels.
- The expertise and facilities required for the investigation, clinical assessment, treatment and holistic management of patients with suspected and established primary immunodeficiencies, autoimmune diseases associated with primary immunodeficiencies and autoinflammatory syndromes.

The service will deliver the aim to improve both life expectancy and quality of life for adults and children with immunodeficiencies by:

- Preventing acute infections or attacks caused by immunodeficiency disorders.
- Halting the progress of complications if present and where possible.
- Reversing previous psychological damage and disability when possible.
- Recognising further complications early and managing them optimally, particularly those not amenable to replacement immunoglobulin therapy.
- Avoiding complications of replacement immunoglobulin therapy.
- Developing approaches to management, based on individual needs, for the lifelong replacement of immunoglobulin, including self administration/home therapy when possible.

## 3.2 Service Description

The provider shall provide hospital-based outpatient and day-care with access to in-patient facilities. This will comprise:

- Regular outpatient clinics for assessment and follow-up.
- Adequate clinical space and staff in relation to the number of patients being treated.
- Development of protocols as required by UK PIN as part of the accreditation process.

- *Central Manchester Foundation Trust: Manchester Royal Infirmary (adults) and Royal Manchester Children's Hospital (children)*

  *Manchester Royal Infirmary (adults):* The immunology department provides a comprehensive clinical immunology, laboratory immunology and clinical allergy service to adults with immunodeficiency within Greater Manchester, the North West of England, North Wales and beyond. The population covered is estimated 3 million.

  The immunology service provides facilities for the investigation, diagnosis and treatment of primary immunodeficiency diseases and allergic diseases. Multidisciplinary team meetings are held with nephrology, rheumatology, ophthalmology and neurology consultants to provide advice on autoimmunity and secondary immunodeficiency. There are two immunology training posts for speciality trainees within the North West deanery that rotate between the clinical services in Salford Royal Foundation Trust and the Manchester Royal Infirmary. The clinical service is staffed by two consultants, three specialist nurses and three members of the administration team.

  *Royal Manchester Children's Hospital*: The Department of Paediatric Allergy and Immunology provides a comprehensive service for the management of primary immunodeficiency to children in the North West. The paediatric population covered is estimated 1.4 million.

  The service provides facilities for the investigation, diagnosis and treatment of primary immunodeficiency diseases and allergic diseases. Multidisciplinary team meetings are held with haematology/BMT, rheumatology, gastroenterology, ENT,

ophthalmology and respiratory consultants. There is one 6 month post for general paediatricians to gain experience in paediatric immunology and allergy within the North West deanery. The clinical service is staffed by two consultants, three specialist nurses and two members of the administration team. The department has close links with the University of Manchester and is active in research into PID.

- *University Hospital of Wales, Cardiff & Vale University Health Board*
  The Department of Immunology at University Hospital of Wales provides a comprehensive clinical Immunology service comprising adult immunology and immunodeficiency, paediatric immunology, the hub diagnostic laboratory for Wales and specialised allergy services. The service also offers home training for immunoglobulin and other immunological therapies. The Department has three Consultant Clinical Immunologists, 3 adult specialist Immunology nurses and 1 paediatric specialist Immunology nurse who are supported by three secretaries in Immunology. The Immunology laboratory comprises immunochemistry, autoimmune, allergy, cellular and molecular sections and supports the investigation, diagnosis and monitoring of the patients seen by the Immunology Department as well as other specialties at UHW, across Wales and internationally.

  There are in addition to the above services specialist clinics for Paediatric Immunodeficiency bone marrow transplantation which are run with the Immunology team in Newcastle, Fever syndrome clinics and the all Wales 22q11.2 MDT clinics.

- *Royal Liverpool & Broadgreen University Hospitals NHS Trust:*
  The Royal Liverpool & Broadgreen University Hospital provides a comprehensive clinical immunology service covering the Merseyside and Cheshire region, including the Isle of Man and North Wales, an approximate population of 3.5 million. Based across 2 sites we diagnose, assess and monitor people who have suspected immunodeficiency due to problems with repeated infections particularly of the chest or sinuses, or severe or unusual infections. We also see patients with acquired or hereditary angioedema due to low C1 esterase inhibitor levels.

  The Trust accepts referrals into the service via fax or letter to the Consultant Immunologist and our outpatient clinic is based

on the Broadgreen Hospital site. The Trust does not currently accept choose and book referral to our clinical immunology clinic. For those patients who require replacement immunoglobulin therapy or acute treatment for their C1 esterase deficiency the immunology specialist nurse manages and maintains a comprehensive home therapy training service and holds a nurse led infusion clinic on the Royal Liverpool site.

The Trust is registered and working towards accreditation with UKPIN and supply data to both the National Immunoglobulin Database and the UKPID Registry. Our service is also an active member of the North West Allergy and Clinical Immunology Network.

## 4. Eligibility Criteria

### 4.1 Eligibility Criteria

**Entry Criteria**

- Any patient suspected of having a PID, hereditary angioedema (HAE) or autoinflammatory condition.

- Immunology PID services primarily involve the diagnosis and management of patients with deficient immune systems, mostly inherited. These are coded for within the World Health Organisation International Classification of Diseases (ICD) codes ICD-10 codes D70, D71, D76, D80-89 (latter group – Certain Disorders involving the Immune Mechanisms).

- Any patient requiring follow up for an immunological disorder that is undefined in the codes above.

**Exit Criteria**

- All patients in whom the above conditions have been excluded.
- End of life
- Where treatment would no longer be appropriate or effective

**Exclusion criteria**

All patients with PID, HAE, and autoinflammatory conditions will require life-long specialist monitoring for recognition and management of complications of disease and therapy. The following exclusion criteria shall apply:

- Patients with HIV-associated immunodeficiency who will be cared for by physicians in Infectious Diseases and GU Medicine.
- Symptoms such as Chronic fatigue syndrome without evidence of immune deficiency.
- Access to the service as described in this specification is for patients with suspected or diagnosed primary immunodeficiency disorders and excludes secondary disorders and the use of IVIg to treat neurological conditions. IVIg for patients with

neurological conditions is typically administered by neurologists on an in-patient (for acute treatment) or day-case basis.

### 4.2 Duty of Care Considerations

The service has a duty of care to ensure that products provided can be used safely by the client and where appropriate carer, personal assistant or support workers.

### 4.3 Expectations of clients and carers

It is a requirement that patients receiving treatment agree to return documentation and regularly attend Immunology OPD appointments for monitoring and safety purposes. This is documented using a formal consent procedure. Failure to comply will result in a warning, then if necessary, removal of home therapy provision.

### 4.4 Palliative Care

Referral to palliative care will be made if required. Care and advice provided by the Immunology team will occur concurrently.

### 4.5 Treatment commenced elsewhere

Patients who have treatment commenced elsewhere, without the benefit of a full immunological work up may need to have treatment stopped to establish baseline information.

## 5. Quality and Patient Safety

### 5.1 Quality and Patient Safety

The Provider must work to agreed written quality standards and provide monitoring information to WHSSC.

The provider must enable the patient's, carer's and advocate's informed participation and to be able to demonstrate this. Provisionwill be made for patients with different information and communication needs for example sensory loss, learning disabilities and for children.

## 5.2 Quality Indicators (Standards)

The Provider must work to the quality standards and provide monitoring information to WHSSC. The quality standards currently issued by UK PIN for accreditation purposes have been adopted in Wales. These are attached at appendix B.

The Providers must be registered and participating in the UK PIN accreditation process.

Providers shall:

- Ensure prescription of therapeutic immunoglobulin will be in accordance with the DH Guidelines (http://www.ivig.nhs.uk).
- Use harmonized patient information and guidelines where available - shared protocols and guidelines have already been developed in professional networks http://www.ukpin.org.uk/home/standards.htm and in some multi-centre regional groups http://www.ukpin.org.uk/home/ to harmonise care and should be used to underpin policy development with patient group involvement.
- Provide a means of collating workload data on inpatient and home therapy workload linked to ICD10 coding including population of a national or local specialist workload monitoring tool.
- Deliver a dashboard for recording outcomes, and more specifically the outcomes related to the key performance indicators as agreed in Wales.
- Work with UKPIN to populate national and international disease registries including the UK PID Registry http://www.ukpin.org.uk/home/registry-introduction.htm.
- Act as ambassadors for the service and support patient and professional organisations improving support and care for conditions under their remit.
- Develop regional care pathways or comply with national care pathways and referral criteria. There is no national care pathway or NICE guidance for PID but one should be developed in collaboration with UK PIN.
- Ensure that Specialist Centre staff support peer accreditation processes if possible by acting as inspectors.
- Support training and education to ensure continuity of future service provision. The provider shall have active participation in training and development of the next generation of specialist clinical immunologists.

- Ensure that Clinical Immunologists will maintain expertise by fulfilling the CPD requirements of the Royal College of Pathologists and/or Physicians/MARS and undertaking team based practice.
- Ensure that the centre has an active role in audit as defined in UKPIN standards (http://www.ukpin.org.uk/home/standards.htm).

## 5.3 Patient Outcomes

Patient outcome measures will be reported to commissioners via an annual audit day.

- The service shall have a CGD antibiotic/management protocol, including antifungal therapy and management protocols for other rare immunodeficiencies under their care as part of their UKPIN Quality Manual.
- The provider should have a policy for ensuring continuity of immunoglobulin supply including ensuring plurality of IVIG/SCIG use to minimize dependence on single supplier.
- The provider shall ensure a patient and public engagement strategy for the service to ensure that patient views of the service are measured (in collaboration with patient organisations).
- The provider shall undertake Patient Related Experience Measures (PREM) surveys for patients and carers on an annual basis.
- The provider shall ensure that there are defined arrangements for maintaining expertise in the management of very rare diseases where there are less than 5 patients per network as well as ensuring the network has sufficient patients to maintain expertise. This may be achieved by ensuring that there are nominated individuals with expertise across the range of very rare disorders per network and through regular educational meetings and through appropriate protocols in the quality manual.
- Unless there is a clear reason not to do so, Clinic letters will be copied to patients.
- Outcome measures will be monitored in line with the UK PIN requirements.
- Decisions that influence the Immunology service provision and financial implications will be made at WHSSC management group level rather than local or directorate level.

## 5.4 Putting Things Right: Raising a Concern

Whilst every effort has been made to ensure that decisions made under this policy are robust and appropriate for the patient group, it is acknowledged that there may be occasions when the patient or their representative are not happy with decisions made or the treatment provided. The patient or their representative should be guided by the clinician, or the member of NHS staff with whom the concern is raised, to the appropriate arrangements for management of their concern:

- When a patient or their representative is unhappy with the decision that the patient does not meet the criteria for treatment further information can be provided demonstrating exceptionality. The request will then be considered by the All Wales Individual Patient Funding Request Panel. The request will normally be made by the patient's GP or Hospital Consultant in writing.

  A patient information leaflet is available via the following link, which explains the process http://www.wales.nhs.uk/sites3/page.cfm?orgid=898&pid=59092. The leaflet also explains how to make a complaint.

- When a patient or their representative is unhappy with the care provided during the treatment or the clinical decision to withdraw treatment provided under this policy, the patient and/or their representative should be guided to the LHB for NHS Putting Things Right http://www.wales.nhs.uk/sites3/home.cfm?orgid=932for services provided outside NHS Wales the patient or their representative should be guided to the NHS Trust Concerns Procedure with a copy of the concern being sent to the Welsh Health Specialised Service Committee.

- If the patient or their representative is not happy with the decision of the All Wales IPFR Panel the patient and/or their representative has a right to ask for this decision to be reviewed. The grounds for the review, which are detailed in the All Wales Policy: Making Decisions on Individual Patient Funding Requests (IPFR) http://www.wales.nhs.uk/sitesplus/documents/867/All%20Wales%20NHS%20IPFR%20Policy.pdf, must be clearly stated. The Local Health Board (LHB) in which the patient is resident should undertake the review;

## 6. Performance Monitoring and Information Requirements

### 6.1 Performance Monitoring

Welsh Health Specialised Services Committee (WHSSC) will be responsible for commissioning services in line with this policy. This will include agreeing appropriate information and procedures to monitor the performance of organisations.

For the services defined in this policy the following approach will be adopted:

- **Service providers to evidence quality and performance controls**
- **Service providers to evidence compliance with standards of care**

WHSSC will conduct performance and quality reviews on an annual basis.

Where there is a single handed Immunologist the provider shall ensure that they actively participate in regional network clinical meetings, to review and compare practice and share expertise in these rare conditions. A minimum attendance requirement at 50% of network meetings (from a total minimum of 4 meeting per annum per network) will be necessary. For rare disease attendance at International meetings is also required.

The provider shall ensure mandatory participation in shared audit across the network. The provider shall ensure that all services in a network share and compare their dashboard performances in a process of continuous quality improvement.

### 6.2 Coding and Activity Monitoring

The provider shall develop an approach to improving the recording and collection of routine activity and performance data.

The provider shall ensure that out-patient as well as in-patient activity for diagnosed patients should be measured using hospital systems to detect patients with the relevant ICD (where one exists). This activity should include the cost of immunoglobulin, C1 inhibitor or other specified high cost drugs unless these are agreed contract exclusions.

There should be a mechanism to collect data on activity related to patients infusing at home. This should include the costs of immunoglobulin, C1 inhibitor, or other high cost drugs, disposables, delivery and nurse time for training and lifelong monitoring.

## 6.3 Accreditation and Quality Standards

All centres should participate and actively work towards United Kingdom Primary Immunodeficiency Network (UKPIN Accreditation) and complete the UKPIN Accreditation Application Form. The accreditation standards are attached at appendix B.

The service should also ensure that:

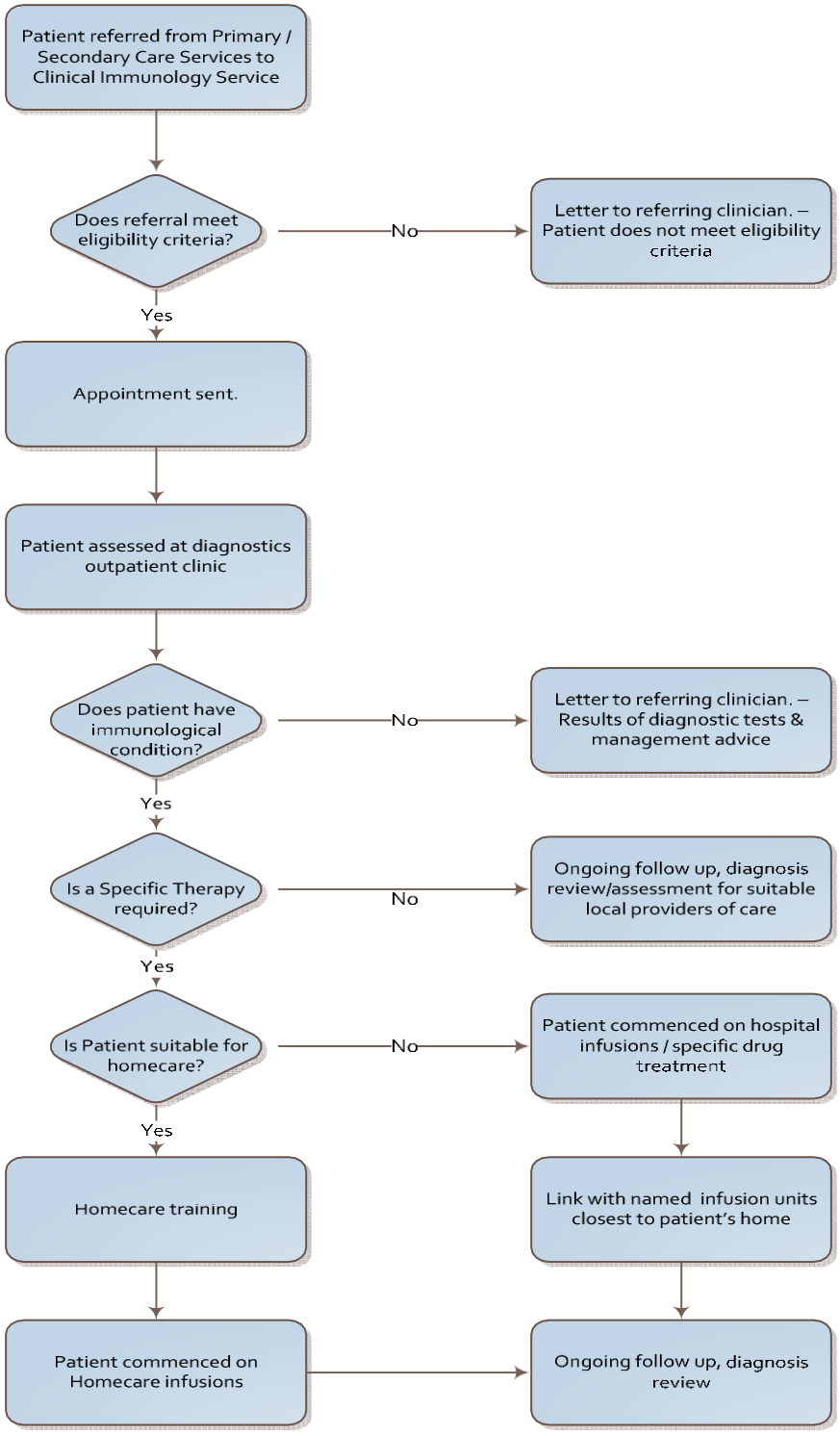
- The management of all patients with any form of primary antibody deficiency should be led by a clinical immunologist with appropriate training and experience and up-to-date CPD.
- Patients should be offered a choice of route (intravenous or subcutaneous) and location (hospital or home) for immunoglobulin replacement therapy if appropriate. All patients should have the opportunity to be assessed for home therapy if appropriate.
- A clinical immunologist should initiate treatment with immunoglobulin, after full risk assessment for that patient and provision of written information.
- Immunoglobulin replacement therapy should be provided by specialist immunology nurses in an established immunology centre and they should be involved in ongoing management of patients receiving therapy both in the home or hospital setting.
- Clinical immunologists should review patients regularly on an outpatient or day-case 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.
- A mechanism to ensure there is documented consent and risk assessment before initiating treatment with blood products including immunoglobulin/C1 inhibitor.
- The provider shall monitor trough or steady state immunoglobulin levels regularly to optimise treatment and review the need for ongoing treatment on an annual basis.

## 6.3 Key Performance Indicators

The providers will be expected to monitor against the agreed key performance indicators for Wales. These are attached at appendix C.

The provider should also monitor the appropriateness of referrals into the service and provide regular feedback to referrers on inappropriate referrals, identifying any trends or potential educational needs.

## Appendix A. Referral Pathway



## Appendix B Quality Indicators (Standards)

The quality standards currently issued by UK PIN for accreditation purposes have been adopted in Wales. A copy of the standards are available via the UK PIN website
http://ukpin.org.uk/home/accreditation-standards.html

## Appendix C. Key Performance Indicators

**KPI 1: To ensure that patients on long term immunoglobulin therapy receive annual reviews.**

| | |
|---|---|
| **Definition**<br>The percentage of patients on long term immunoglobulin therapy who receive annual reviews, including clinical letter to their General Practitioner. | |
| **Rationale** | Annual reviews are necessary to ensure that patients are receiving the appropriate level of immunoglobulin therapy. |
| **Assessment Criteria** | Number of patients on long term immunoglobulin therapy, and percentage who have received an annual review over the last 12 months. |
| **Target** | 90% of patients to receive an annual review |
| **Data Source** | To be recorded by the specialised immunology services. |
| **Validation** | Annual audit of a subset of patients |
| **Reporting Arrangements** | To be reported annually to the WHSSC Information Team. |

**KPI 2: To ensure that patients on long term immunoglobulin therapy receive at least 6 monthly trough level measurements.**

| | |
|---|---|
| **Definition**<br>The percentage of patients on long term immunoglobulin therapy who receive 6 monthly trough level measurements. | |
| **Rationale** | Six monthly trough level measurements are necessary to ensure that patients are receiving appropriate management. |
| **Assessment Criteria** | Number of patients on long term immunoglobulin therapy, and percentage who have received six monthly trough level measurements over the last 12 months |
| **Target** | 90% of patients to receive six monthly trough level measurements. |
| **Data Source** | To be recorded by the specialised immunology services. |
| **Validation** | Annual audit of a subset of patients |
| **Reporting Arrangements** | To be reported annually to the WHSSC Information Team. |

**KPI 3: To monitor the informed consent process for immunoglobulin and C1 esterase inhibitor therapy.**

| | |
|---|---|
| **Definition**<br>Patients receiving immunoglobulin and C1 esterase inhibitor therapy require informed consent which is documented in the medical notes. | |
| **Rationale** | To monitor the compliance with this standard. |
| **Assessment Criteria** | Evidence of informed consent in the notes. |
| **Target** | 100% informed consent in the notes. |
| **Data Source** | To be recorded by the specialised immunology services. |
| **Validation** | Annual audit of a subset of notes. |
| **Reporting Arrangements** | To be reported annually to the WHSSC Information Team. |

**KPI 4: To monitor the quality of laboratory diagnostic testing.**

| | |
|---|---|
| **Definition**<br>Patients being investigated for immunodeficiency require laboratory testing in accredited laboratories. | |
| **Rationale** | To monitor the compliance with this standard. |
| **Assessment Criteria** | Evidence of certificates of CPA accreditation for the Immunology, Haematology and Biochemistry laboratories where testing is performed. |
| **Target** | Evidence of CPA Accreditation Certificates for all of the above laboratories where testing is performed. |
| **Data Source** | CPA Certificates from the Laboratory Quality Manager for the above Laboratories. |
| **Validation** | Up to date CPA Accreditation Certificates. |
| **Reporting Arrangements** | To be reported annually to the WHSSC Information Team. |