GIG CYMRU NHS WALES
Cyd-bwyllgor Comisiynu
Joint Commissioning Committee

Specialised Immunology

Service Specification: SS78

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Description	NHS Wales will routinely commission this specialised service in accordance with the criteria described in this policy
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Document Update Information

The policy includes updated terminology.

There have been no changes to the patient pathway.

GIG CYMRU NHS WALES | Cyd-bwyllgor Comisiynu Joint Commissioning Committee

Contents

Statement

NHS Wales Joint Commissioning Committee (NWJCC) will commission specialised immunology services in accordance with the criteria outlined in this specification.

In creating this document NWJCC has reviewed the requirements and standards of care that are expected to deliver this service.

Welsh Language

NWJCC is committed to treating the English and Welsh languages on the basis of equality, and endeavour to ensure commissioned services meet the requirements of the legislative framework for Welsh Language, including the Welsh Language Act (1993), the Welsh Language (Wales) Measure 2011 and the Welsh Language Standards (No.7) Regulations 2018.

Where a service is provided in a private facility or in a hospital outside of Wales, the provisions of the Welsh language standards do not directly apply but in recognition of its importance to the patient experience, the referring health board should ensure that wherever possible patients have access to their preferred language.

In order to facilitate this, NWJCC is committed to working closely with providers to ensure that in the absence of a Welsh speaker, written information will be offered and people have access to either a translator or 'Language-line' if requested. Where possible, links to local teams should be maintained during the period of care.

Decarbonisation

NWJCC is committed to taking assertive action to reducing the carbon footprint through mindful commissioning activities. Where possible and taking into account each individual patient's needs, services are provided closer to home, including via digital and virtual access, with a delivery chain for service provision and associated capital that reflects the NWJCC commitment.

Disclaimer

NWJCC assumes that healthcare professionals will use their clinical judgment, knowledge and expertise when deciding whether it is appropriate to apply this document.

This document may not be clinically appropriate for use in all situations and does not override the responsibility of healthcare professionals to make decisions appropriate to the circumstances of the individual patient, in consultation with the patient and/or their carer or guardian, or Local Authority.

NWJCC disclaims any responsibility for damages arising out of the use or non-use of this policy.

1. Introduction

This document has been developed as the Service Specification for the planning and delivery of specialised immunology services for people of all ages who are affected by inborn errors of immunity and are resident in Wales. This service will only be commissioned by the NHS Wales Joint Commissioning Committee (NWJCC) and applies to residents of all seven Health Boards in Wales.

1.1 Background

Specialised Immunology services are for patients with inborn errors of immunity.

Inborn errors of immunity (IEI) are rare diseases that occur when components of the immune system are either not present or not functioning normally. They arise due to changes in specific genes that encode proteins involved in the functioning of the immune system. IEI present clinically as increased susceptibility to infections, autoimmunity, auto-inflammatory diseases, allergy, bone marrow failure, and/or malignancy. IEI can be diagnosed throughout the lifetime of a person, with the most severe forms usually being diagnosed in childhood[1].

Early diagnosis and long-term management are critical to optimise outcomes for patients with IEI. The initial work up of patients with suspected inborn errors of immunity often requires complex immunopathological and genetic investigations to establish a definitive diagnosis. The management of patients with inborn errors of immunity requires either regular, life-long therapy with an expensive blood product with limited availability (immunoglobulin (Ig)), other biological agents such as C1-inhibitor concentrate, monoclonal antibodies, other biologicals, small molecule inhibitors or complex procedures such as bone marrow or thymic transplantation. Use of high cost-low volume drugs including biological agents in complex cases, often through individual funding requests is frequently required. Management involves multidisciplinary teams consisting of Specialist Doctors, Specialist Nurses, Dieticians, Physiotherapy, Psychologist and Social work support.

Epidemiology

The 2022 Update on the Classification of Human Inborn Errors of Immunity[2] from the International Union of Immunological Societies[3] (IUIS) Expert Committee reports a total of 485 inborn errors of immunity, adding 55 novel monogenic gene defects and one

[1] http://www.immunodeficiencyuk.org/
[2] Human Inborn Errors of Immunity: 2022 Update on the Classification from the International Union of Immunological Societies Expert Committee, Journal of Clinical Immunology, 42, pp1473–1507, June 2022
[3] https://iuis.org/

phenocopy due to autoantibodies since the previous update, published in January 2020. Ten categories of IEI are listed by the IUIS:

- combined immunodeficiencies
- combined immunodeficiencies with syndromic features
- predominantly antibody deficiencies
- diseases of immune dysregulation
- congenital defects of phagocytes
- defects in intrinsic and innate immunity
- autoinflammatory diseases
- complement deficiencies
- bone marrow failure
- phenocopies of inborn errors of immunity

While individually rare, in aggregate, the prevalence of these conditions is approximately 1 in 1000 to 5 in 1000 meaning there would be up to 15,500 people in Wales affected by an IEI. About 20 IEI account for over 90 per cent of cases meaning that many conditions are extremely rare[4].

1.2 Aims and Objectives

The aim of this service specification is to define the requirements and standard of care essential for delivering specialised immunology services for people with IEI.

The objectives of this service specification are to:

- detail the specifications required to deliver specialised immunology services for people who are residents in Wales
- ensure minimum standards of care are set for specialised immunology services
- ensure equitable access to specialised immunology services
- identify centres that are able to provide specialised immunology services for Welsh patients
- improve outcomes for people accessing specialised immunology services

[4] Immunodeficiency UK

1.3 Relationship with other documents

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

- **NHS Wales**
  - All Wales Policy: Making Decisions in Individual Patient Funding requests (IPFR).

- **Welsh Government**
  - The Transition and Handover Guidance, February 2022

- **National Institute of Health and Care Excellence (NICE) guidance**
  - Transition from children's to adults' services for young people using health or social care services, NG43, 2016

- **Other published documents**
  - QPID standards for diagnosis and management of PID
  - British Society for Immunology Competency Framework for Immunology Nursing

2. Service Delivery

The NHS Wales Joint Commissioning Committee will commission the specialised immunology service for patients of all ages with inborn errors of immunity (IEI) in line with the criteria identified in this specification.

2.1 Access Criteria

The specialised immunology service is for patients of all ages having, or suspected of having, an IEI.

2.2 Service description

In addition to the standards required within the Contract, specific quality standards and measures will be expected. The provider must also meet the standards as set out below.

Specialist Immunology centres should provide:

- A high quality, accessible and sustainable service that meets the needs of the local population, reflects effective resource use and incorporates the views of patients.
- Excellent, holistic, multidisciplinary care for patients with IEI according to best practice guidelines defined by the British Society for Immunology Clinical Immunology Professional Network (BSI-CPIN, formerly UKPIN), European Society for Immunodeficiencies (ESID) and other authoritative bodies.
- The expertise and facilities required for the investigation, clinical assessment, treatment and holistic management of patients with suspected and established IEI.
- Equity of access to best practice standards, based on current guidelines for diagnosis and management for patients with IEI and related complications.
- Integrated care with primary, secondary and other care providers and ensure close links and collaboration with other expert centres at national and international levels.
- Training for future specialists to maintain service continuity.

Facilities and equipment

Clinic space

Hospital-based outpatient and day-care with access to in-patient facilities should be provided according to standards set by regulation and accreditation bodies. This should 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.
- Access to in-patient beds should be available and admission pathways for patients with IEI should be established with individualised care plans where necessary.

Diagnostics

A diagnostic package comprised of routine and complex tests for the investigation of suspected immunodeficiency should be delivered, 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 genetic NGS +/- tissue typing.
- Specialised Immunology Laboratory services with UKAS accreditation or equivalent.
- Access to diagnostics for rare and emerging diseases through UK/European/USA laboratories.

Pharmacy

The service should have appropriate pharmacy 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 storage facilities* for blood product immunological therapies and to allow reliable traceability documentation of dispensing to individual patients.

*Immunoglobulin and C1 esterase inhibitor may be dispensed by blood bank or pharmacy but should have enhanced batch tracking systems in place which meet Medicines and Healthcare Products Regulatory Agency (MHRA), accreditation and blood regulatory requirements.

Specialist teams

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

- An appropriate number of Consultant Clinical Immunologists or equivalent with experience in management of patients with IEI and who maintain up-to-date Continuing Professional Development (CPD) in their area of practice for the caseload and in order to provide cover;

- An appropriate number of Senior Specialist Nurses with immunology experience and training for the caseload to provide nursing care, training and run the home treatment service.
- Physiotherapist(s)

Self-Care

Self-care should be provided as an option based on the patient's wishes, abilities and circumstances and should include:

- Provision of information about when to seek advice from the specialist 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, biologicals, immunological therapies or immunoglobulin therapy IgRT (intravenous or subcutaneous, or facilitated), infusion sets, pumps for subcutaneous delivery, deliveries of consumables to patients' homes, regular outpatient consultations and monitoring of immune status, antibody levels, blood counts and liver function tests.

Home care programmes should be accredited under the Quality in Primary Immunodeficiency Services (QPIDS) programme.

Patients with confirmed IEI requiring regular immunoglobulin replacement therapy should 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 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 drugs) for treatment or prophylaxis (e.g. surgical, dental or investigational procedures), long term prophylaxis including managing those patients on home therapy.
- Management of those immunodeficiencies requiring other/new treatments (e.g. monoclonal antibodies, biologicals, immunological therapies or cytokines) on a named patient basis, where there is a suitable evidence base. This includes day case attendance, nursing supervision, the drug, pumps for subcutaneous or intravenous use, home therapy options, monitoring by biochemical tests, specialised immunopathological tests and medical follow-up.

Specialist Clinics

The specialised immunology service should offer or have links to the following specialist clinics which should involve appropriate expertise and take place at appropriate frequency:

- Bone marrow transplant clinic
- Paediatric Fever/Auto inflammatory syndrome clinic
- 22q11.2 MDT clinic
- Fever/Autoinflammatory clinic

The service should triage and identify patients requiring referral to highly specialised national services for specialist procedures such as haematopoietic stem cell transplant (HSCT) or thymic transplant.

2.3 Interdependencies with other services or providers

The service should have timely access to related services required for the optimal care of immunodeficiency patients. This should include access to psychologists, dieticians, social workers and play therapy. It also includes access to optimal therapeutic and diagnostic vaccination.

The service should have timely access to support from other clinical specialties (both adult and paediatric) for complications of IEI including:

- Cardiology
- Ear, Nose and Throat Medicine
- Respiratory Medicine
- Gastroenterology
- Dermatology
- Infectious Diseases
- Haematology Transplant
- Haematology/Oncology
- Neurology
- Ophthalmology
- Psychiatry
- Paediatrics
- Paediatric and Neonatal Intensive care
- Clinical Genetics
- Rheumatology
- Psychology
- Nephrology

- Microbiology
- Virology

The provider should maintain the following links:

Secondary care links

Depending on the nature of the immune disease, services should be involved in shared care in relation to general medical needs, delivery of antibiotics and, for some patients, immunoglobulin therapy. 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.

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 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.

Primary care links

Care plans of IEI patients should be shared with primary care.

Antibiotic guidelines should be shared with general practitioners.

Home therapy and management should be arranged in liaison with primary care.

Clinic letters should be sent to GPs and other specialties involved in a patient's care and the patient or parent.

Private sector and third sector links

The service should maintain strong liaison with appropriate Primary Immunodeficiency Patient Groups and signpost patients to appropriate support organisations.

The provider should ensure Home therapy delivery services are available which may be contracted out to third party suppliers for delivery agency of immunoglobulin C1 inhibitor concentrate and other medicinal products to patients' homes as required.

2.4 Transition arrangements

All children and young people with an IEI need a coordinated transitional care programme leading to transfer of care to an adult immunology service at an appropriate age, under a shared care arrangement.

All transition arrangements should be in line with Transition from children's to adults' services for young people using health or social care services NICE guidance NG43 and Welsh Government guidance on transition and handover from children's to adult health services.

Transition involves a process of preparation for young people and their families for their transition to adulthood and their transition to adult services. This preparation should start from early adolescence 12-13 year olds. The exact timing of this will ideally be dependent on the wishes of the young person but will need to comply with local resources and arrangements.

The transition process should be a flexible and collaborative process involving the young person and their family as appropriate and the service with the support of the third sector where relevant.

Shared protocols between child and adult services should be established, defining the roles and responsibilities of each member of the teams.

2.5 Exclusion Criteria

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 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.

Note that NWJCC has not been delegated routine commissioning of services for patients with secondary immunodeficiency. Funding will be considered on an individual basis via the process outlined in 2.9 below.

2.6 Acceptance Criteria

The service outlined in this specification is for patients ordinarily resident in Wales, or otherwise the commissioning responsibility of the NHS in Wales. This excludes patients who whilst resident in Wales, are registered with a GP practice in England, but includes patients resident in England who are registered with a GP Practice in Wales.

2.7 Patient Pathway (Annex i)

Referrals can be made from both primary and secondary care[5] as follows:

- Due to the complex nature of IEI, tertiary referrals into the immunology services come from Tier 2 (general physicians) or other Tier 3 tertiary or specialist physicians (particularly respiratory, ENT, rheumatology, 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.

2.8 Designated Centres

Cardiff & Vale University Health Board
University Hospital of Wales
Heath Park
Cardiff
CF14 4XW

Manchester University NHS Foundation Trust
Manchester Royal Infirmary
Oxford Road
Manchester
M13 9WL

[5] Newborn bloodspot screening currently being piloted in England. If adopted in Wales, some patients will be identified and referred to the service via the screening pathway.

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 (PSCID) • BMT • Gene therapy • Autoimmune Gut Diseases (AGD) • Juvenile Idiopathic Arthritis(JIA)
Newcastle upon Tyne Hospitals NHS Foundation Trust	• Paediatric Severe Combined Immunodeficiency and Related Disorders (SCID) • BMT • Autoimmune Gut Diseases (AGD) • Juvenile Idiopathic Arthritis(JIA) • Specialist lab testing
Royal Free Hampstead NHS Trust	• Cryopyrin Associated Periodic • BMT • Specialist lab testing
University Hospital Bristol NHS Foundation Trust	• Barth Syndrome • Transplants for Paediatrics

2.9 Exceptions

If the patient does not meet the criteria for treatment as outlined in this policy, an Individual Patient Funding Request (IPFR) can be submitted for consideration in line with the All Wales Policy: Making Decisions on Individual Patient Funding Requests. The request will then be considered by the All Wales IPFR Panel.

If the patient wishes to be referred to a provider outside of the agreed pathway, an IPFR should be submitted.

Further information on making IPFR requests can be found at: Individual Patient Funding Requests

3. Quality and Patient Safety

The provider must work to written quality standards and provide monitoring information to the lead commissioner. The quality management systems must be externally audited and accredited.

The centre must enable the patients, carers and advocates informed participation and to be able to demonstrate this. Provision should be made for patients with communication difficulties and for children, teenagers and young adults.

3.1 Aims of the Service

The service will 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 IgRT and other biological or immunological therapies.
- Developing approaches to management, based on individual needs, for the lifelong replacement of immunoglobulin, including self-administration/home therapy when possible.
- Developing approaches to management, based on individual needs, for the lifelong management of hereditary angioedema (HAE) for acute treatment, pre-procedure prophylaxis and prophylactic therapies.
- Assessment, management and follow-up of patients requiring HSCT or thymic transplantation.

3.2 Quality Indicators (Standards)

The service should:

- work closely with the Immunoglobulin Strategy Group for Wales which manages Ig supply in Wales
- have a Chronic Granulomatous Disorder (CGD) antibiotic management protocol, including antifungal therapy and management protocols for other rare immunodeficiencies under their care as part of their QPIDs Quality Manual.
- have a patient and public engagement strategy for the service to ensure that patient views of the service are measured (in collaboration with patient organisations).

- undertake Patient Related Experience Measures (PREM) surveys for patients and carers on an annual basis.
- ensure that there are defined arrangements for maintaining expertise in the management of very rare diseases where there are fewer 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.
- attend Regional Immunology Network meetings.
- use harmonised patient information and guidelines where available - shared protocols and guidelines have already been developed in professional networks and in some multi-centre regional groups 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 outcomes related to the key performance indicators specified in section 4.3.
- work with BSI-CIPN to populate national and international disease registries including the UK PID Registry
- 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.
- 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 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.
- ensure that specialist nurses are either compliant with, or working towards, the British Society for Immunology Competency Framework for Immunology Nursing
- ensure that 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.
- ensure that a clinical immunologist should initiate treatment with immunoglobulin, after full risk assessment for that patient and provision of written information.
- ensure that IgRT 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.

- ensure that 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.
- ensure that a mechanism is in place to ensure there is documented consent and risk assessment before initiating treatment with blood products including immunoglobulin/C1 inhibitor.
- monitor trough or steady state immunoglobulin levels regularly to optimise treatment and review the need for ongoing treatment on an annual basis.

3.3 National Standards

Providers should be registered and participating in the Quality in Primary Immunodeficiency Services (QPIDS) accreditation process. Outcome measures will be monitored in line with the UK PIN requirements.

3.4 Other quality requirements

- The provider will have a recognised system to demonstrate service quality and standards.
- The service will have detailed clinical protocols setting out nationally (and local where appropriate) recognised good practice for each treatment site.
- The quality system and its treatment protocols will be subject to regular clinical and management audit.
- The provider is required to undertake regular patient surveys and develop and implement an action plan based on findings.

4. Performance Monitoring and Information Requirement

4.1 Performance Monitoring

NWJCC 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

NWJCC will conduct performance and quality reviews on an annual basis

The provider should 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 should ensure mandatory participation in shared audit across the network.

4.2 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.

4.3 Key Performance Indicators

The providers will be expected to monitor against the full list of Quality Indicators derived from the service description components described in Section 2.2.

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.

In particular, the provider will be expected to monitor against the following target outcomes which apply to both the adult and paediatric service:

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

Definition 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 service.
Validation	Annual audit of a subset of patients.
Reporting Arrangements	To be reported annually to the NWJCC Information Team.

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

Definition 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 NWJCC Information Team.

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

Definition 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 NWJCC Information Team.

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

Definition 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 UKAS accreditation for the Immunology, Haematology and Biochemistry laboratories where testing is performed.
Target	Evidence of UKAS Accreditation Certificates for all of the above laboratories where testing is performed.
Data Source	UKAS Certificates from the Laboratory Quality Manager for the above Laboratories.
Validation	Up to date UKAS Accreditation Certificates.
Reporting Arrangements	To be reported annually to the NWJCC Information Team.

KPI 5: To monitor the number of adverse events documented on the data base for patients on long term immunoglobulin.

Definition The number of moderate to severe adverse events per 1000 treatment days for patients on long term immunoglobulin. Moderate to severe adverse effects are defined as systemic allergic reactions (anaphylaxis), thrombosis, haemolysis, aseptic meningitis, renal failure, significant dermatological adverse effects and viral transmission.	
Rationale	To monitor the quality of service delivered by the specialised immunology centres.
Assessment Criteria	Number of adverse events per 1000 treatment days.
Target	0.01 adverse event per 1000 treatment days.
Data Source	To be recorded by the specialised immunology services.
Validation	Any serious adverse outcomes that result in patient harm should be reported to NWJCC with learning from these. Themes, trends and numbers should be included.
Reporting Arrangements	To be escalated within the Health Board Quality and Patient Safety framework and reported to NWJCC through these channels.

KPI 6: To monitor the auditing of deaths whilst under the care of the specialised immunology service.

Definition

All deaths in patients to be audited, referred to relevant clinical governance committee and mortality review.	
Rationale	To monitor the deaths relating to patients receiving service delivered by the specialised immunology centres.
Assessment Criteria	Total number of deaths in last year, total number of deaths reported to clinical governance committee and also findings of audits relating to deaths.
Target	To audit all deaths in the relevant period.
Data Source	To be recorded by the specialised immunology services.
Validation	Minutes of mortality review meeting and audit findings reported to commissioners.
Reporting Arrangements	Feedback and review from trends and themes emerging from mortality and morbidity reviews on a quarterly basis. Any death that occurs as an outcome of service delivery should be reported immediately in line with Quality and safety standards through the Health Board and to Commissioners.

KPI 7: To monitor patient experience whilst under the care of the specialised immunology service.

Definition A validated patient experience measure is required for monitoring of patients perspectives of service delivered.	
Rationale	Patient experience is a useful measure of a systems / holistic view of the service and also can herald problems with service delivery
Assessment Criteria	Use of the CAREs patient experience tool to monitor and measure patient experience both for the unit as a whole. For further information see www.caremeasure.org
Target	All patients admitted or seen in outpatients in the last year
Data Source	Questionnaire responses to be recorded by the specialised immunology services.
Validation	Audit findings to be presented to commissioners at review meetings or annual audit meetings
Reporting Arrangements	To be reported annually to the NWJCC Information Team.

KPI 8: To monitor quality of life outcome measures patient experience whilst under the care of the specialised immunology service.

Definition A validated Quality of Life measure is required for monitoring of patients perspectives of service delivered	
Rationale	Quality of Life Measures are a fundamental approach to measuring patient outcome measures.
Assessment Criteria	An annual audit of EQ-5D, SF-6D or similar changes before and after treatment is commenced 6 months after commencing therapy.
Target	All patients admitted or seen in outpatients in the last year.
Data Source	Questionnaire responses to be recorded by the specialised immunology services.
Validation	Audit findings to be presented to commissioners at review meetings or annual audit meetings.
Reporting Arrangements	To be reported annually to the NWJCC Information Team.

KPI 9: To ensure that patients with immunodeficiency receive appropriate multidisciplinary team support.

Definition The percentage of immune deficient patients who have access to specialist MDT members, including specialist doctors, specialist nurses, dietitians, physiotherapists, psychologists and social care support.	
Rationale	Appropriate review by specialist healthcare professionals is necessary to ensure that patients have complex care needs assessed.
Assessment Criteria	Number of patients identified as requiring additional support and the percentage who have received such an annual review with that specialist 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 NWJCC Information Team.

4.4 Date of Review

This document is scheduled for review before 2027 where we will check if any new evidence is available.

If an update is carried out the policy will remain extant until the revised policy is published.

5. Equality Impact and Assessment

The Equality Impact Assessment (EQIA) process has been developed to help promote fair and equal treatment in the delivery of health services. It aims to enable NHS Wales Joint Commissioning Committee to identify and eliminate detrimental treatment caused by the adverse impact of health service policies upon groups and individuals for reasons of race, gender re-assignment, disability, sex, sexual orientation, age, religion and belief, marriage and civil partnership, pregnancy and maternity and language (Welsh).

This policy has been subjected to an Equality Impact Assessment.

*The Assessment demonstrates the policy is robust and there is no potential for discrimination or adverse impact. All opportunities to promote equality have been taken.

6. Putting Things Right

6.1 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.

If 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. For 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 NWJCC.

6.2 Individual Patient Funding Request (IPFR)

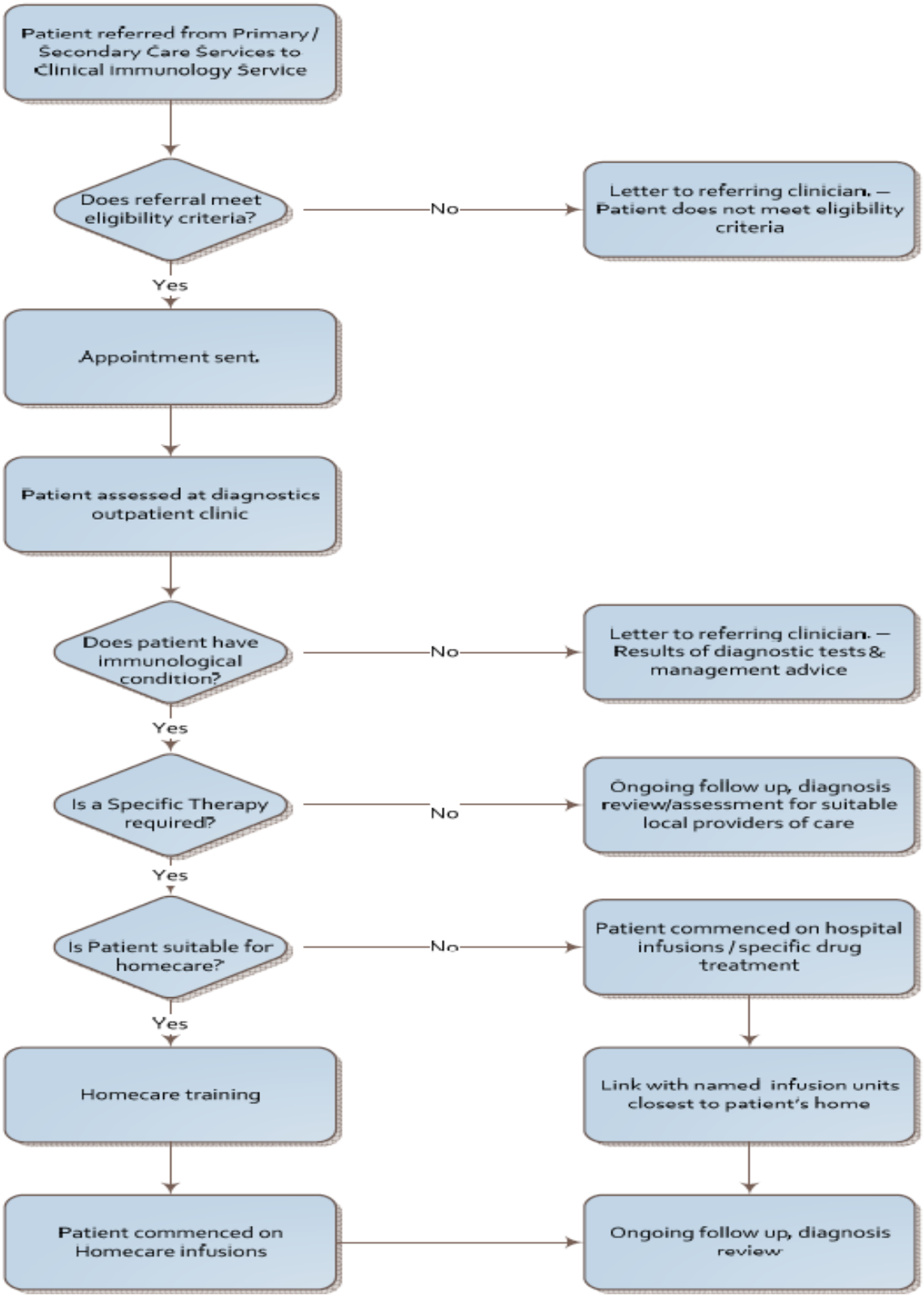
If the patient does not meet the criteria for treatment as outlined in this policy, an Individual Patient Funding Request (IPFR) can be submitted for consideration in line with the All Wales Policy: Making Decisions on Individual Patient Funding Requests. The request will then be considered by the All Wales IPFR Panel.

If an IPFR is declined by the Panel, a patient and/or their NHS clinician has the right to request information about how the decision was reached. If the patient and their NHS clinician feel the process has not been followed in accordance with this policy, arrangements can be made for an independent review of the process to be undertaken by the patient's Local Health Board. The ground for the review, which are detailed in the All Wales Policy: Making Decisions on Individual Patient Funding Requests (IPFR), must be clearly stated.

If the patient wishes to be referred to a provider outside of the agreed pathway, and IPFR should be submitted.

Further information on making IPFR requests can be found at: Individual Patient Funding Requests

Annex i Patient Pathway



Annex ii Codes

Code Category	Code	Description
ICD 10	D70	Agranulocytosis
	D71	Functional disorders of polymorphonuclear neutophils
	D76	Other specified diseases with participation of lymphoreticular and reticulohistiocytic tissue
	D80-89	Certain disorders involving the immune mechanism

Annex iii Abbreviations and Glossary

Abbreviations

AGD	Autoimmune Gut Diseases
BMT	Bone Marrow Transplant
BSI-CPIN	British Society for Immunology Clinical Immunology Professional Network
ESID	European Society for Immunodeficiencies
CGD	Chronic Granulomatous Disorder
CPD	Continuing Professional Development
HAE	Hereditary Angioedema
HIV	Human immunodeficiency virus
HSCT	Haematopoietic stem cell transplantation
IEI	Inborn Errors of Immunity
Ig	Immunoglobulin
IgRT	Immunoglobulin Replacement Therapy
IPFR	Individual Patient Funding Request
IUIS	International Union of Immunological Societies
IVIg	Intravenous Immunoglobulin
JIA	Juvenile Idiopathic Arthritis
MHRA	Medicines and Healthcare Products Regulatory Agency
NGS	Next Generation Sequencing
NICE	National Institute for Health and Care Excellence
NWJCC	NHS Wales Joint Commissioning Committee
QPIDS	Quality in Primary Immunodeficiency Services
SCID	Severe Combined Immunodeficiencies
SCIg	Subcutaneous Immunoglobulin
PSCID	Paediatric Severe Combined Immunodeficiency and Related Disorders
UKAS	United Kingdom Accreditation Service
UKPIN	United Kingdom Primary Immunodeficiency Network

Glossary

Individual Patient Funding Request (IPFR)

An IPFR is a request to NHS Wales Joint Commissioning Committee (NWJCC) to fund an intervention, device or treatment for patients that fall outside the range of services and treatments routinely provided across Wales.

NHS Wales Joint Commissioning Committee (NWJCC)

NWJCC is a joint committee of the seven local health boards in Wales. The purpose of NWJCC is to ensure that the population of Wales has fair and equitable access to the full range of Tertiary Services. NWJCC ensures that services within our portfolio are commissioned from providers that have the appropriate experience and expertise. They ensure that these providers are able to provide a robust, high quality and sustainable services, which are safe for patients and are cost effective for NHS Wales.