B09/S/a

2013/14 NHS STANDARD CONTRACT
FOR SPECIALISED IMMUNOLOGY (ALL AGES)

SECTION B PART 1 - SERVICE SPECIFICATIONS

Service Specification No.	B09/S/a
Service	Specialised Immunology (All Ages)
Commissioner Lead	
Provider Lead	
Period	12 months
Date of Review	

1. Population Needs

1.1 National/local context and evidence base

Primary immunodeficiency disorders (PID) are a group of primarily genetic disorders of the immune system in which part of the immune system is missing or does not function properly. Primary denotes the mainly genetic nature of the defects, differentiating them from secondary or acquired immunodeficiencies caused by malnutrition, infection (e.g., human immunodeficiency virus [HIV] infection), chemotherapy or other external agents. Immunology PID services primarily involve the diagnosis and management of patients with deficient immune systems, mostly inherited (mainly included within ICD-10 codes D70, D71, D76, D80-89 (latter group – Certain Disorders involving the Immune Mechanisms).

There are over 150 different but rare immune deficiencies recognized by the European Society for Immunodeficiencies (www.esid.org), IUIS classification 2011 (http://www.iuisonline.org/iuis/index.php/primary-immunodeficiency-expert-committee.html), Herz W et al. Frontiers in Immunology 2011;2:54), of which less than 20 account for > 90% of cases (see table 1). These diseases range in prevalence from 1 in 3,000 to less than one in a million (overall estimate 1/15,000). The rarity, wide spectrum, severity of complications and associated mortality and complexity of treatments require that PID be managed by immunology specialists (UKPIN Consensus http://www.ukpin.org.uk/home/managers.html, UKPIN standards of care for CVID and SCID http://www.ukpin.org.uk/home/standards.htm). A brief overview of the more common conditions can be found on the UKPIN website http://www.ukpin.org.uk/home/PIA-archive/1-questions.htm.

Table 1: Main Primary Immunodeficiency Disorders (PID)
Common variable immunodeficiency disorders (CVID)
X-linked agammaglobulinaemia (XLA)
Specific antibody deficiency
IgG subclass deficiency
IgA deficiency
Hyper-IgE syndrome (HIGE)
Hyper-IgM syndromes (HIGM)
X-linked lymphoproliferative syndromes
Other humoral immunodeficiencies
Wiskott-Aldrich syndrome (WAS)
Ataxia telangiectasia (AT)
22q11 deletion syndromes (Di George)
Chronic mucocutaneous candidiasis (CMC)
Other T cell immunodeficiencies
IFN/IL-12 deficiencies
Severe Combined Immunodeficiencies (SCID) (all forms)
Chronic Granulomatous Disorder (CGD)
Other neutrophil disorders
C1 inhibitor deficiency (Hereditary & Acquired Angioedema (HAE & AAE)
Other complement component deficiencies

Because PIDs are relatively rare, they are frequently overlooked and reports in the North West area and nationally have highlighted a diagnostic delay of up to 10 years before immunoglobulin replacement therapy is started (Seymour et al. Primary antibody deficiency and diagnostic delay. J Clin Path 2005;58:546-7). The delay is longer in adult patients with antibody deficiency, paediatricians being more aware of the possibility of PIDs than those working in adult medicine. The delay in diagnosis contributes to increased morbidity and mortality in this patient group: overall, 25 year survival from diagnosis in this group of patients is around 75% compared to 92% for the population of similar age, but 50% of those with complications die in a similar timeframe. (European CVID Database, Chapel et al. Blood 2008;112; 277-86). Unrecognised PID contributes to increased morbidity and mortality and the mean diagnostic delay in Europe is 7.5 years. About half of the patients without a diagnosis will be admitted to hospital every year; they may also be seen in different specialty clinics for a range of complications. They often receive almost continuous antibiotics for infections and are off work for long periods of time. Delayed diagnosis remains a concern for physicians and patients and continues to be an issue (Consensus document: Primary antibody deficiencies: recognition, clinical diagnosis and referral of patients. Clinical Medicine. 2009;9(6);595-599, http://www.rcplondon.ac.uk/resources/concise-guidelines-primary-antibody-deficiencies and patient evidence from Hereditary Angioedema (HAE) UK http://www.haeuk.org/patient-stories/ and the Chronic Granulomatous Disorder Society (CGD Society)(Jones LB et al. Chronic granulomatous disease in the UK and Ireland:a comprehensive national patient-based registry. Clin Exp Immunol 2008;152:211-8). Specific advice for commissioners on PID commissioning has been produced by UKPIN http://www.ukpin.org.uk/home/managers.html, as has an integrated care pathway incorporating patient stories.

In total there are probably up to 5000 Primary immune deficient (PID) patients in England (adults and children combined) including C1inhibitor deficiency. Currently 1,950 patients listed on the UKPID registry database from 2012. Many of these centres have just commenced data entry and none have completed the process. There are an additional 11 centres yet to commence data entry. The national immunoglobulin demand management database reports 2,078 patients on long-term intravenous and subcutaneous immunoglobulin replacement for antibody deficiency as of March 2012 in England. Table 2 provides an estimated prevalence of the sub categories of PID assuming a total prevalence of 5000 patients in England.

Table 2: Prevalence data for immunodeficiencies in England	
Category	**Estimated number of patients**
Primary antibody deficiency	3,018
Other well defined PID	455
Complement deficiency	435
Primarily T cell disorders	432
Unclassified PID	256
Neutrophil phagocytic disorders	108
Autoimmune and immune dysregulation syndromes	28
Defects of innate immunity	20
Autoinflammatory disorders	0

* PID based on current representation of diagnoses in patients already on the UKPID Registry and based on international registry below (Toplak et al.)
There are approximately 600-900 patients with Hereditary Angioedema (HAE) on active treatment, 500 patients under investigation or observation but not on treatment, and 500-1000 with other primary immunodeficiencies not requiring immunoglobulin replacement in the UK as a whole. There may be some additional patients on immunoglobulin by home delivery which are not yet captured as of March 2012, but will be in future.

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Immunology Services also provide diagnosis and management of rare autoinflammatory syndroms (Cryopyrinopathies, TRAPS and others) involving immunomodulatory biological therapies as referenced in National Specialist Services Definitions Set (NSSDS) 17. These are rare, newly described conditions(see Table 3) in which there is abnormal multisystem inflammation, associated with deficiencies in the Innate immune system. They can present to any specialty with organ specific inflammation, and are often under-recognised. Many individual with these conditions have identifiable genetic associations, which these enable targeting of specific biologic drugs. The majority of patients with these conditions are likelyto be under the care of Immunology services, and are included in specialised rheumatology services or in collaboration with rheumatologists. http://www.specialisedservices.nhs.uk/library/26/Specialised_Immunology_services_all_ages.pdf. A recent international registry documented 228 patients with autoinflammatory disorders in the UK (Toplak et al. An international registry on autoinflammatory diseases: the Eurofever experience. Ann Rheum Dis 2012; 71: 1177-1182).

Table 3 - Major autoinflammatory conditions of known genetic aetiology
(adapted from Lachmann HJ. An approach to the patient with a periodic fever syndrome. Clin Exp Immunol 2011;165:301-309)

- Familial Mediterranean Fever (FMF)
- Tumour necrosis factor receptor-associated periodic syndrome (TRAPS)
- Mevalonate kinase deficiency (Hyper-IgD syndrome)
- Cryopyrin-associated periodic syndromes (CAPS):
- Familial cold autoinflammatory syndrome (FCAS)
- Muckle-Wells syndrome
- Neonatal onset multisystem inflammatory disease (NOMID)
- PAPA – pyogenic arthritis, pyoderma gangrenosum and acne
- Blau syndrome
- DIRA – deficiency of interleukin-1-receptor antagonist

What is involved in the management of immunological disorders?
The initial work up of patients with suspected PID or autoinflammatory syndromes often requires complex immunopathological and genetic investigations to establish a definitive diagnosis (see service inter-dependencies and diagnostic support).
The management of patients with established primary immune deficiencies or autoinflammatory syndromes 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 or complex procedures such as bone marrow 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.

Severe immunodeficiencies typically require bone marrow transplantation (SCID, other T cell deficiencies, CD40 ligand deficiency, X-linked lymphoproliferative syndrome, Wiskott-Aldrich syndrome, CGD, IFN/Il-12 deficiencies, other neutrophil disorders). These are provided for children in the two supraregional services in

London and Newcastle. Post-transplant care however is usually undertaken in collaboration with Specialist PID centres.

Antibody disorders require life-long replacement antibody replacement (CVID, XLA, some specific antibody deficiency) and continual specialist monitoring for complications, including local care of post-transplant patients shared with the national transplant services. A minority of secondary immunodeficiencies are sufficiently severe to require management in the same way as a primary immunodeficiency and these patients would also require Specialist care in these centres and may require immunoglobulin.

Complement disorders require access to replacement complement factors (C1 inhibitor deficiency (hereditary and acquired angioedema) and continual specialist monitoring for complications.

Other immune disorders have complex medical needs (Hyper-IgE syndrome, Ataxia telangiectasia, 22q11 deletion syndromes, CMC) and require and continual specialist monitoring for complications and collaborative management with other services for non-immune complications.

National Context: The Existing Services

There are approximately 26 centres in England in 2012. These will mostly be registered on the British Society for Immunology Clinic list “find an immunologist” http://www.immunology.org/page.aspx?pid=1349 and/or on the BSACI list for their allergy services or listed on the UKPIN list of centres www.ukpin.org.uk.

A national professional organization, The UK Primary Immunodeficiency Network (UKPIN), exists to develop and disseminate national standards of care, maintain centre registration, maintain the UKPID Registry and to provide a structured framework for accreditation. The UK has a network of specialist immunology services which cover most but not all of England in a geographically equitable distribution (UKPIN) and should be embedded in new commissioning structures. All centres are expected to participate in national registry data collection, training, examination, peer inspection and guideline development for UKPIN and research into PID as an orphan disease and this should be included in any national commissioning strategy. A peer-review accreditation process with professionally agreed Service standards and an inspection process have been developed through the UKPIN professional network to harmonise care http://www.ukpin.org.uk/home/accreditation-standards.html .

The UKPIN accreditation scheme for PID centres was set up in 2000 following the findings of a Department of Health-sponsored national audit of PID services. The intention of the scheme is to improve the care of PID patients through common approaches to diagnosis, care, support and management by means of setting consensus standards of practice and service organisation. Following an initial pilot phase the accreditation scheme became established in 2007 under the direction of a Network Accreditation Committee. Within the scheme, PID services are evaluated against agreed and ratified standards which are re-evaluated and updated as

necessary by UK PIN. The process involves an initial self-assessment of local service compliance with the standards followed by formal application for accreditation and external peer review through consideration of information supplied in the application by the Accreditation Committee and a site assessment visit by professional specialty assessor.

Home Care Programs:

Approximately half of all immunoglobulin replacement for PID is delivered through managed home care programmes involving a multidisciplinary team.

Services for children and specialised transition arrangements:

Adult and paediatric services are usually separate but work closely together and are often co-located. Transition arrangements for seamless transfer of care between adult and paediatric services should be in place in specialist centres but formal arrangements are probably lacking in many at present.

Care for patients with autoimmune complications associated with immunodeficiency syndromes as defined by European guidelines is also provided by existing services (these patients are a subset of the PID above). Specialist immunological management of complex autoimmune and vasculitic conditions, including diagnosis and treatment is often undertaken in collaboration with rheumatology, respiratory, neurology and other organ-based specialist services in small subsets of patients;
(http://www.rcplondon.ac.uk/resources/publications/consultant-physicians-working-patients).

National Policies of relevance:

Long term conditions

http://www.dh.gov.uk/prod_consum_dh/groups/dh_digitalassets/@dh/@en/documents/digitalasset/dh_4122574.pdf.

Rare diseases

http://www.dh.gov.uk/en/Publicationsandstatistics/Publications/PublicationsPolicyAndGuidance/DH_132880.

Networks

http://healthandcare.dh.gov.uk/clinical-senates-and-networks/.

Immunoglobulin demand management plan

http ://www.ivig.nhs.uk/

Equity and Excellence

http://www.dh.gov.uk/prod_consum_dh/groups/dh_digitalassets/@dh/@en/@ps/documents/digitalasset/dh_117794.pdf.

Transition:

Guidelines for best practice in providing transitional services for young people as they move from paediatric to adult services have been developed and will be a key component of an adult and paediatric specialist service.

http://www.dh.gov.uk/en/Publicationsandstatistics/Publications/PublicationsPolicyAndGuidance/DH_083592.

Accreditation:

UK PIN standards for diagnosis and management of PID and position statements are available at: http://www.ukpin.org.uk/home/standards-position-statement.htm
and http://www.ukpin.org.uk/home/standards.htm

Key additional evidence and guidance on the investigation and management of antibody deficiency and C1 inhibitor deficiency are as follows:

Consensus document Primary antibody deficiencies: recognition, clinical diagnosis and referral of patients. Clinical Medicine. 2009;9(6);595-599, http://www.rcplondon.ac.uk/resources/concise-guidelines-primary-antibody-deficiencies.

Bonilla FA et al.American Academy of Allergy, Asthma and Immunology Practice Parameter for the diagnosis and management of primary immunodeficiency. Ann Allergy Asthma Immunol 2005;94:S1-S63.

Diagnostic guidance has been adapted with referral guidance by professional organisations and patient organisations which can be expanded and promulgated more widely;

http://www.isitpid.com/
http://www.cgdsociety.org/medicalprofessionals/generalmedicalinformation/diagnostictoolchecklist
http://www.ukpin.org.uk/home/ESID/index.htm

Gompels, MM and Lock, RJ and Abinun, M and Bethune, CA and Davies, G and Grattan, C and Fay, AC and Longhurst, HJ and Morrison, L and Price, A and Price, M and D Watters, D. 2005. C1 inhibitor deficiency: consensus document. Clinical and Experimental Immunology , 139 (3) 379 – 394.

Cicardi M et al. Evidence-based recommendations for the therapeutic management

of angioedema owing to C1 inhibitor deficiency; consensus report of an international working group. Allergy 2012;67:147-157. A consensus report on behalf of the Hereditary Angioedema Working Group (HAWK).

2. Scope

2.1 Aims and objectives of service

The provider shall ensure that Specialist Immunology centres will provide:

- A high quality, accessible and sustainable service that meets the needs of the local population and reflects effective resource use and incorporates the views of patients.
- Excellent, holistic, multidisciplinary care for patients with immunodeficiency, complex autoimmunity and auto-inflammatory syndromes according to best practice guidelines defined by UKPIN, 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 primary immunodeficiencies, autoimmune diseases associated with primary immunodeficiencies and autoinflammatory syndromes.
- Equity of access to best practice standards, based on current guidelines for diagnosis and management for patients with PID 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.Train future specialists to maintain service continuity.

The service will deliver the aim to improve both life expectancy and quality of life for adults 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.

2.2 Service description/care pathway

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 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 Biologic and Cytotoxic infusion facilities. This service should be supported by clear guidelines, protocols, and pathways for patient care, in which are embedded the key principles of Chemotherapy safety, As outlined in the document “Chemotherapy Services in England: Ensuring quality and safety. A report from the National Chemotherapy Advisory Group August 2009” adopted where appropriate for use in non-cancer chemotherapy.

The provider shall have access to support from other clinical specialties for complications of PID including:

- Ear, Nose and Throat Medicine, Respiratory Medicine, Gastroenterology, Infectious Diseases, Haematology/ Oncology, Paediatrics, Clinical Genetics, Rheumatology.

The provider 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, test immunisations, specialised genetic and radiology studies: Specifically this will require:

- 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 provider shall have appropriate pharmacy facilities including:

- Appropriate storage and dispensing facilities for drugs and immunoglobulin products.
- Pharmacy storage facilities for immunological therapies and good documentation of dispensing to individual patients.
- Pharmacy support for maintenance of the Immunoglobulin demand management programme database.

The provider 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.
- The provider shall ensure that all home care programmes should be accredited

through UKPIN.

- 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 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 immunodeficiencies requiring other/new treatments (e.g. monoclonal antibodies 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, monitoring by biochemical tests, specialised immunopathological tests and medical follow-up.

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

- At least two Consultant Clinical Immunologists or equivalent with experience in management of patients with PID and who maintain up-to-date CPD in their area of practice (see comments above on single-handed practice).
- Senior Specialist Nurses with immunology experience and training to provide nursing care, training and run the home treatment service.

The provider shall provide transition services:

- For children with PID before referral to adult services based on the framework recommended by the Department of Health.www.dh.gov.uk/transition Children with PID are transferred to the adult service between the ages of 16 and 18 years.
- Transfer arrangements and preferences should be discussed with the child and their family up to 12 months in advance. Shared protocols between child and adult services should be established, defining the roles and responsibilities of each member of the teams.

The provider shall maintain the following links:

- Secondary care links
    - Depending on the nature of the immune disease, services are involved in shared care in relation to general medical needs, delivery of antibiotics and, for some patients, immunoglobulin therapy (small number of patients receive Ig therapy at peripheral hospitals) with:
- Primary care links
    - Care plans of PID patients are shared with primary care.
    - Antibiotic guidelines are shared with general practitioners.
    - Home therapy and management is arranged in liaison with CCGs.

    - 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 Primary Immunodeficiency Patient Groups – including the Chronic Granulomatous Disorder Society, Hereditay 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.

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

Referral processes and sources

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 to ensure the referral requires specialist input. A care pathway with referral guidance should be developed.

Equity of access to services

No patient should have to travel excessively for access to local expert centres. Patients with rarer diseases requiring referral to a national specialist centre or centres should have equitable access and distance to travel wherever possible, taking account of geographical issues.

Some centres provide specialist services to other health economies (Wales, Scotland, NI, Republic of Ireland).

Location(s) of Service Delivery

- All current centres in England as above.

Days/Hours of operation

- The provider shall ensure that services are available during office hours.
- The provider shall ensure that there is a written agreed patient pathways for dealing with out of hours emergencies and a system for giving out-of-hours advice. These will include antibiotics for infections and C1 inhibitor or Icatibant for angioedema attacks in HAE and admission policies for PID.

Response time & detail and prioritisation

- As per national waiting time targets.

Service user/ carer information

- The provider shall ensure that centres will provide (in collaboration with patient organisations where they exist):
  - written disease-specific information leaflets.
  - periodic educational events for patients.
  - periodic educational events for GPs.
  - information to patients and staff about patient support organisations.
- The provider shall ensure that Specialist Centre Staff support patient groups with membership of Medical Advisory panels.

Entry Criterion

Any patient suspected of having a PID or autoinflammatory condition as detailed above.

Exit Criterion

All patients in whom the above conditions have been excluded.

All patients with PID and autoinflammatory conditions will require life-long specialist monitoring for recognition and management of complications of disease and therapy.

2.3 Population covered

The service outlined in this specification is for patients ordinarily resident in England*; or otherwise the commissioning responsibility of the NHS in England (as defined in Who Pays?: Establishing the responsible commissioner and other Department of Health guidance relating to patients entitled to NHS care or exempt from charges).

*Note: for the purposes of commissioning health services, this EXCLUDES patients who, whilst resident in England, are registered with a GP Practice in Wales, but INCLUDES patients resident in Wales who are registered with a GP Practice in England.

Specifically, this service is for adults with immunodeficiency syndromes requiring specialised intervention and management, as outlined within this specification. The provider will ensure that:

- Each hub centre will operate in a network with approximately 250 PID patients to maintain sufficient expertise.

Each centre develops a regional patient pathway for access to PID services which ensures that only patients with suspected immune deficiency or HAE or associated complications are referred. This can be supported with web-based referral decision tools such as the ESID/UKPIN diagnostic algorithm.

http://www.ukpin.org.uk/home/ESID/index.htm

2.4 Any acceptance and 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.

2.5 Interdependencies with other services

The provider 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 and ophthalmology and behavioural medicine.

The provider shall deliver close input from physiotherapy - essential for the management of the respiratory complications associated with PID.

The provider shall ensure access to social workers, psychologists and dieticians for selected patients completes the package of holistic care required for PID patients.

For centres without paediatric immunologists, the provider shall triage and identify patients requiring referral to highly specialised national services for immunodeficiency at GOS and Newcastle= and their collaboration Paediatric Immunology Specialist Services

The provider shall deliver access to close support from a high-quality, accredited diagnostic immunology laboratory providing a range of routine and specialist assays.

While most services do not have access to in-patient beds, admission pathways for PID patients should be established with individualized care plans where necessary.

The provider will work collaboratively with Paediatric Specialist Immunology and Infectious Diseases Services Specification.

3. Applicable Service Standards

3.1 Applicable national standards e.g. NICE, Royal College

The provider shall ensure that centres achieve UKPIN accreditation. Currently there are 5 fully accredited centres in England with the others registering or preparing for accreditation. This process shall eventually be mandatory for Specialist centres.

The provider shall ensure that Centres shall be active members and participants of UKPIN as evidenced by:

- UKPIN (http://www.ukpin.org.uk/home/standards.htm), the European Society for Immunodeficiencies (ESID)
- The International Working Group on C1 inhibitor deficiency (Cicardi et al. Allergy 2012;67: 147-57) Other authoritative bodies such as the Royal College of Pathologists and the Royal College of Physicians and listed in section 1.
- UK PIN standards for diagnosis and management of PID and position statements are available at: http://www.ukpin.org.uk/home/standards-position-statement.htm and http://www.ukpin.org.uk/home/standards.htm.
- The provider shall ensure that prescription of therapeutic immunoglobulin will be in accordance with and monitored by the DH Guidelines and Demand Management Programme (http://www.ivig.nhs.uk). In some Trusts, immunology services are also responsible for provision of high-dose immunoglobulin therapy for patients with autoimmune conditions e.g. neuropathies.
- The provider shall 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.
- The provider shall 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 (web-based database).
- The provider shall deliver a dashboard for recording outcomes, process or proxy measures.
- The provider shall deliver a means for populating national and international disease registries including the UK PID Registry http://www.ukpin.org.uk/home/registry-introduction.htm.
- The provider shall act as ambassadors for the service and support patient and professional organisations improving support and care for conditions under their remit.
- The provider shall 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.
- The provider shall ensure that Specialist Centre staff support peer accreditation processes by acting as inspectors.
- The provider shall 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.
- The provider shall ensure that Clinical Immunologists will maintain expertise by fulfilling the CPD requirements of the Royal College of Pathologists and/or Physicians and undertaking team based practice.
- The provider will ensure that the centre has an active role in audit as defined in UKPIN standards (http://www.ukpin.org.uk/home/standards.htm).

4. Key Service Outcomes

Expected Outcomes - Adult Clinical

Immunology

Patient Outcomes

- The provider 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 organizations). The provider shall undertake Patient Related Experience Measures (PREM) surveys for patients and carers on an annual basis and achieve >75% satisfaction and act on any deficiencies identified.
- The provider shall comply with the requirements of the Immunoglobulin Demand Management Plan.
- 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.
- Outcome measures will be monitored in accordance with the Consensus document Primary antibody deficiencies: recognition, clinical diagnosis and referral of patients. Clinical Medicine. 2009;9(6);595-599, http://www.rcplondon.ac.uk/resources/concise-guidelines-primary-antibody-deficiencies. Specifically:
  - All patients with a symptomatic proven primary antibody deficiency and/or structural organ damage should receive immunoglobulin replacement therapy.
  - All patients with a primary antibody deficiency should be monitored regularly for the occurrence of acute infection. The provider shall ensure documentation of number of infections requiring admission and/or antibiotics each year.
  - All patients with a primary antibody deficiency should be monitored regularly for development of disease progression and complications.
  - Unless there is a clear reason not to do so, Clinic letters will be copied to patients.

Clinical governance

- 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.
- 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.

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.

Accreditation and Quality Standards:

- All centres should participate and actively work towards UKPIN Accreditation and complete the UKPIN Accreditation Application Form.

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 immmunoglobulin levels regularly to optimise treatment and review the need for ongoing treatment on an annual basis.

5. Location of Provider Premises

Specialist Clinical Immunology services are currently provided from 26 centres in England as listed by UKPIN.