

A14/S(HSS)/C

**2013/14 NHS STANDARD CONTRACT  
FOR LYMPHANGIOLEIOMYOMATOSIS SERVICE (ADULTS)**

**PARTICULARS, SCHEDULE 2 – THE SERVICES, A - SERVICE  
SPECIFICATIONS**

<b>Service Specification No.</b>	A14/S(HSS)/c
<b>Service</b>	Lymphangiomyomatosis service (adults)
<b>Commissioner Lead</b>	
<b>Provider Lead</b>	
<b>Period</b>	12 months
<b>Date of Review</b>	

## 1. Population Needs

Lymphangiomyomatosis (LAM) is a rare, progressive disease categorised by lung cysts, kidney tumours and lymphatic abnormalities. LAM can occur in isolation or as part of the genetic disease tuberous sclerosis (TSC). Patients have repeated lung collapse and respiratory impairment leading to death or lung transplantation and they can also suffer complications from kidney tumours.

LAM is a heterogeneous disease with a variable clinical phenotype. The majority of patients with symptomatic disease will experience a decline in lung function and disability increasing over a variable period of years. A smaller number of patients with the disease have few symptoms but require initial assessment and the employment of strategies to minimise disease progression. A smaller number of patients have highly aggressive disease and progress to respiratory failure within five years. In a previous study performed to examine disability in the UK it was concluded that in 10 years from diagnosis half of patients will be too breathless to walk on the flat, 25% will be using supplemental oxygen, 10% will be housebound due to breathlessness and a similar number will have died of the disease (Johnson et al. thorax 2004).

### 1.1 National/local context and evidence base

Lymphangiomyomatosis (LAM) is a rare, progressive disease categorised by lung cysts, kidney tumours and lymphatic abnormalities.

The aim of the service is for a clinic to provide diagnosis, medical management and

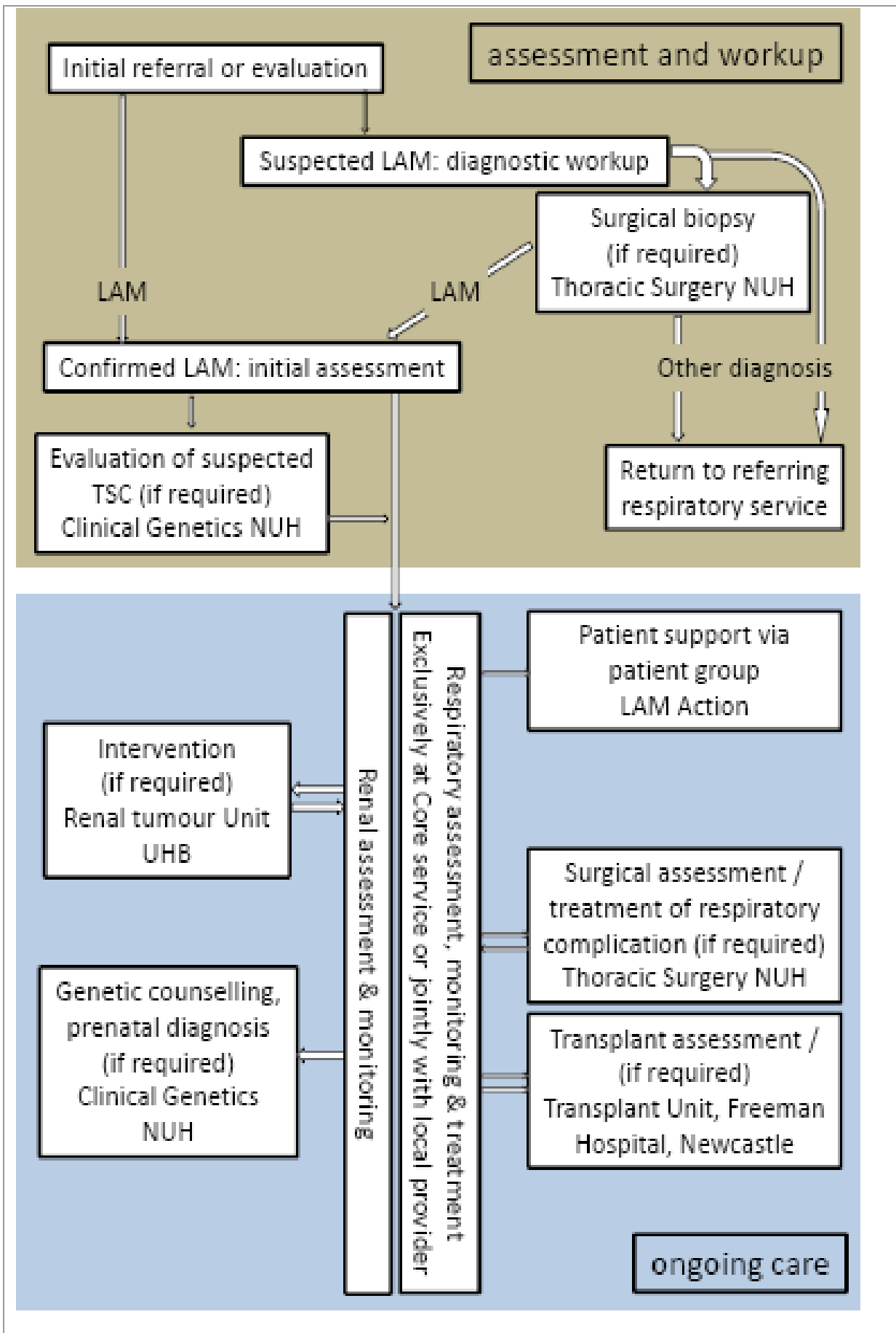
care co-ordination for patients with LAM.

Expert management of LAM is likely to improve quality of life for patients and may slow the decline in lung function and progression to lung transplant. Co-ordination of surgery for lung and kidney problems reduces morbidity and improves quality of life.

Treatment of LAM involves identifying and optimising a series of related conditions in multiple systems. Appropriate identification and treatment of these particular problems reduces morbidity, increases quality of life and reduces health service costs. Aspects of treatment which are frequently managed inappropriately are discussed below.

### **Treatment of pneumothorax**

70% of patients have pneumothorax, and this is recurrent in two thirds of these (Johnson et al. Thorax 2000). On average patients have over four pneumothoraces each of which results the need for chest drainage and an average hospital stay of eight days per episode (Al Moosa et al. Chest 2008). Appropriate treatment at the first pneumothorax is recommended in current European Respiratory Society Guidelines, significantly reduces recurrence rates in LAM minimises morbidity and reduces hospital in patient stay costs and the complications of chest drain insertion (Johnson and Tattersfield Thorax 2000, Johnson et al. European Respiratory Journal (ERJ) 2010, Al Moosa et al. Chest 2008).



## **2. Scope**

### **2.1 Aims and objectives of service**

- to make a definite diagnosis where appropriate using the least invasive means and in keeping with recent clinical guidelines (Johnson et al. ERJ 2010);
- to identify tuberous sclerosis where present in patients with LAM resulting in appropriate targeted treatment and genetic counselling;
- to provide multi-disciplinary (lung, thoracic surgical, kidney, transplant, tuberous sclerosis complex (TSC)) evaluation and clinical care for LAM to newly diagnosed and existing patients: either as sole provider or through shared care with local health services;
- to improve technical expertise, the timing of transplant referral and optimise communication over assessment and follow up of these patients;
- to ensure all patients receive optimal evidenced based care appropriate to their stage of disease and in keeping with international guidelines (Johnson et al. ERJ 2010);
- to ensure appropriate use of mTOR inhibitors to prevent loss of lung function in patients likely to benefit from this therapy;
- to ensure easy access to specialist advice for patients and other health care providers.

### **2.2 Service description/care pathway**

#### **Delivery of care**

The LAM service provides a comprehensive service for patients with monitoring run from the core at Nottingham University Hospitals NHS Trust (NUH). Patients found not to have LAM after initial workup by the centre will be returned to their referring service. Others will have continuing monitoring and specified care by the core service. This will be performed exclusively by the core service or in conjunction with their local health care provider according to the patient's wishes and practical considerations including travel.

Patients will be seen at other parts of the service as determined by clinical need according to predetermined protocols.

#### **Days/hours of operation**

This is a 24-hour, 7-day a week service.

#### **Outpatient assessment and management:**

This will be delivered as 30 x 4 hour clinics/year by the lead consultant physician and the respiratory nurse specialist. The predicted workloads are shown below (section

8).

**In patient care:**

For management of complications and in some cases diagnostic workup it is estimated that 10 patients/year (with a mean stay of seven days each) will require inpatient treatment and/or investigation. Patients will be admitted by the provider under the care of the lead consultant physician.

**Evaluation of patients with TSC and suspected TSC - Clinical Genetics NUH:**

Based upon an audit of our experience to date it is expected that five patients per year will be referred to the Department of Clinical Genetics at NUH for further diagnostic workup of suspected TSC including genotyping. In addition a further two patients may require other assessments including pre-natal diagnosis during pregnancy.

**Surgical treatment - Thoracic Surgery NUH:**

When clinically indicated video assisted thoroscopic lung biopsy will be utilised. Liaison with surgical colleagues will also be required for effective management of pneumothorax and pleural effusion.

**Management of renal angiomyolipoma - Urology Unit, University Hospitals Birmingham NHS Foundation Trust:**

The LAM service will liaise with the Urology Unit at Queen Elizabeth Hospital, University Hospitals Birmingham (UHB) NHS Foundation Trust in order to provide appropriate expert care for patients with renal angiomyolipoma. UHB NHS Foundation Trust will provide case discussions and surgical procedures as clinically required under sub-contract arrangements with Nottingham University Hospitals NHS Trust.

**Lung transplant referral - Cardiopulmonary Transplant Unit, The Newcastle upon Tyne Hospitals NHS Foundation Trust:**

When clinically indicated the LAM service will refer appropriate patients to nationally designated lung transplant service at The Newcastle upon Tyne Hospitals NHS Foundation Trust.

**Drug treatment:**

Patients with rapidly progressive disease or specific complications for which no other therapy is available will be considered for treatment with the mTOR inhibitor Rapamycin. Patients likely to benefit from rapamycin according to current evidence are those with rapid deterioration in lung function, those with chylous complications and those with complex angiomyolipomas not amenable to renal surgery. A protocol for patient selection and monitoring of rapamycin therapy has been produced.

### **Patient review, administration and liaison with other providers:**

Coordination of investigations, writing reports, and planning treatment in conjunction with other providers e.g. University Hospitals Birmingham NHS Foundation Trust Urology unit or The Newcastle upon Tyne Hospitals NHS Foundation Trust Transplant Unit, will be a major portion of the workload. This is estimated at 42 consultant sessions/year and time from the service coordinator.

Patients will, from time to time, also require management of complications of LAM with their local healthcare providers. The LAM service will liaise with these providers whenever necessary to provide support and expert advice but the service is not responsible for these, non-highly specialised, elements of the patients overall care.

### **Discharge criteria**

The National Centre for LAM, together with local providers aims to deliver comprehensive care for LAM patients at all stages of their disease.

Patients discharged from the service will be those found not to have LAM after their initial evaluation (see flow chart in section 3). These patients will be returned to their original referring care provider.

### **Risk management**

Care delivered by the LAM service provider must be of a nature and quality to meet the care standards, specification and agreement for the service. It is the trust's responsibility to notify the commissioner on an exceptional basis should there be any breaches of the care standards. Where there are breaches any consequences will be deemed as being the trust's responsibility.

Patients must be managed in line with the specification and care standards. Any deviation from these which has not been approved by the NHS England is at the trust's risk both clinically and financially. It is the trust's responsibility to inform the commissioners of any such non-approved deviations on an exceptional basis.

Where a patient's presentation challenges the assumptions that underpin the specification, service standards and contractual arrangements it is the trust's responsibility to inform the commissioners on an exceptional basis, prior to any treatment (except for emergency treatment) so that the implications of the patient's requirements can be considered. This does not affect situations where the Individual Funding Application process applies.

The service will liaise closely with LAM Action, the UK LAM patient group. Patients will provide patient oriented outcome measures and a patient satisfaction survey will be completed and acted upon after each year of service.

The provider will work with NHS England to ensure sufficient considerations are given to communications.

## **2.3 Population covered**

This service covers patients registered with an English General Practitioner, resident in Scotland, resident in the European Union and eligible for treatment in the NHS under reciprocal arrangements.

Patients from Wales and Northern Ireland are not part of this commissioned service and the trust must ensure it has separate arrangements in place.

## **2.4 Any acceptance and exclusion criteria**

The centre will review patients with suspected LAM for diagnostic purposes or to plan on-going care in those with established disease. Where the diagnosis of LAM is confirmed baseline evaluation and specified on-going care will be offered. Referrals will be accepted from any clinician in the relevant area. Self referrals from patients will not be accepted.

### **Minimum referral criteria:**

Patients will be evaluated at the centre in the presence of any of the following:

- a definite or possible diagnosis of LAM1 made at any centre
- a thoracic CT-scan showing diffuse cystic lung disease not attributable to an existing disease in women with or without respiratory symptoms
- respiratory disease in patients with tuberous sclerosis complex
- respiratory disease in patients with a history of renal angiomyolipoma.

The centre will review radiology and discuss referral with the treating clinician for any patients with diffuse cystic lung diseases in any context.

All relevant investigations performed by referring centres (including radiology, biopsies, lung function tests) will be reviewed to avoid duplication of investigations.

### **Evaluation of clinical referrals:**

All referrals and accompanying data will be reviewed by the LAM centre clinical team. Where further information or the results of investigations are required, these will be sought prior to the patient's evaluation at the centre.

### **Exclusion criteria**

Patients whose referral information does not support the possible diagnosis of LAM.

### **Response time & detail and prioritisation**

The service will meet national waiting time guidelines and targets.

In addition we will aim to:

- read all referrals and clinical questions and where clinically required, respond within one working day or if routine within one week
- evaluate patients at the centre within one month of receiving the appropriate referral and clinical data
- make referrals to other parts of the service within one week of evaluation.

The service is accessible to all patients with LAM who fit the referral criteria and regardless of sex, race, or gender.

The service's staff must attend their trust's mandatory training on equality and diversity and the facilities provided offer appropriate disabled access for patients, family and carers.

When required the service will use translators and printed information is available in multiple languages.

The provider has a duty to co-operate with the commissioner in undertaking Equality Impact Assessments as a requirement of race, gender, sexual orientation, religion and disability equality legislation.

## **2.5 Interdependencies with other services**

Patients may require different aspects of the service for different aspects or stages of their disease. The majority of care will be delivered by NUH with referrals for specific aspects of care made to the other elements of the service or other healthcare providers where appropriate.

**Evaluation and management of renal disease** - Patients with angiomyolipoma require monitoring of these tumours by imaging and may need prophylactic treatment to prevent haemorrhage resulting in emergency nephrectomy. An appropriate schedule of initial evaluation and monitoring will be performed by the core service at NUH in line with current recommendations. Patients requiring interventional treatment according to clinical protocols will be assessed by the renal tumour service at University Hospitals Birmingham.

**Respiratory complications** - Respiratory complications include pneumothorax and chylous pleural effusions. Both of these problems are frequently recurrent and are a cause of significant morbidity for patients with LAM. These complications often require definitive surgical treatment at an appropriate point. This requires liaison between thoracic surgeons and physicians which will occur with the thoracic surgical department in NUH.

**Patient support** - For rare diseases is important, particularly around the time of diagnosis and at other times in patients with advanced disease or prior to lung transplantation. Good links between clinical services, specialist nurse support and patient's groups will facilitate this.



### 3. Applicable Service Standards

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

The nationally designated LAM service will be integrated into the trust's corporate and clinical governance arrangements.

The commissioners and service will conduct a formal Joint Service Review at least every six months.

See also NHS England Service Standards for the Lymphangiomyomatosis (LAM) Service

### 4. Key Service Outcomes

<b>Quality Performance Indicator</b>	<b>Threshold</b>	<b>Method of measurement</b>	<b>Consequence of breach</b>	<b>Report Due</b>
Individual rate of decline of Forced expiratory volume in 1 second (FEV <sub>1</sub> )	>150 ml/yr	serial lung function		
<b>Number of pneumothoraces</b>	>1 without intervention	clinical record		
Incidence of renal haemorrhage	>1 after intervention	clinical record		
Overall survival	less than national average for LAM	clinical record		

### 5. Location of Provider Premises

Nottingham University Hospitals NHS Trust  
QMC campus  
Derby Road  
Nottingham  
NG7 2UH

#### Sub-contractors

In addition to the core service the National LAM referral Centre will also comprise:

University Hospitals Birmingham NHS Foundation Trust  
PO Box 9551  
Mindelsohn Way  
Queen Elizabeth Medical Centre  
Birmingham  
B15 2PR

Adopted April 2013