



# *Papworth Clinic for Adults with Ataxia-Telangiectasia*

*The Ataxia-Telangiectasia Society  
IACR-Rothamsted  
Harpenden, Herts  
AL5 2JQ*

*Tel: 01582 760733  
Fax: 01582 760162*

*email: [atsociety@btconnect.com](mailto:atsociety@btconnect.com)  
website: [www.atsociety.org.uk](http://www.atsociety.org.uk)*

*Reg. Charity No. 1105528*

## Background

In October 2004 the A-T Society organised an international meeting in Birmingham to highlight progress in A-T research and the challenges ahead. An international consensus emerged that chest problems in A-T were a major cause of ill health and death.

Separate studies indicate that a number of factors may contribute to chest problems in A-T including difficulties in swallowing, breathing, and neuromuscular control, infections and a deficient or poorly regulated immune system.

Establishing a programme to assess these factors was recognised as a priority in A-T, with the ultimate aim being to improve quality of life and length of life. Papworth Hospital was identified as the UK centre with the blend of expertise required.

The adult Ataxia-Telangiectasia service at Papworth Hospital includes:

- A detailed respiratory assessment
- An immunological assessment

As well as functional assessments with a:

- Respiratory Physiotherapist
- Occupational Therapist
- Speech and Language Therapist
- Dietitian
- Social Worker.

## Papworth Hospital

The hospital opened in 1917 as a tuberculosis hospital and when Pendrill Varrier-Jones conceived the idea, he wanted to create not just a hospital, but a community. This sense of community remains.

During the 1950s, Papworth became one of the region's main hospitals, developing first thoracic surgery, then cardiac surgery and cardiology. Today, Papworth provides specialised heart and lung services to adults and is the country's main heart and lung transplant centre. Over 20,000 inpatients and day cases and 20,000 outpatients are treated each year, and there are 1,300 staff, and 225 beds.

Papworth Hospital is one of the foremost hospitals in the UK and has all the latest equipment that is needed to assess patients with chest problems and infections.

It is located around 10 miles outside Cambridge with good road access, although public transport is limited.

## Patient Pathway for the A-T Service

**Two** visits to Papworth are required for an initial complete consultation.

The **first** visit is for 2-3 days as an NHS inpatient, and the **second** visit, about 6 weeks later, includes an overnight stay, some follow-up tests and a consultation with feedback and advice about the results to patients and their family/carer as appropriate.

Patients will be offered an **annual review visit**. This entails an overnight stay for some of the tests to be repeated to assess your progress and to see whether any changes to your treatment are needed.

Patients referred to the Adult A-T Clinic at Papworth Hospital will be contacted and offered a date for admission. An information pack with a request for a preliminary blood sample will be sent to you.

**The A-T Society will pay any reasonable costs of transferring you from your home to Papworth and back, together with a member of your family or a carer, if necessary.**

## Respiratory Support and Sleep Centre (RSSC)

This is a specialist unit with purpose-built accommodation. It is here that adults with A-T will be staying.

The RSSC treats large numbers of patients with sleep disorders but also receives referrals from all over the country for patients with conditions affecting their respiratory system. Many patients have chronic disorders of the nervous system which control the pattern of breathing of the breathing muscles themselves. RSSC staff are used to dealing with patients with disabilities due to complex neurological disorders and the implications of these for their activities of daily living.

Their aim is to personalise the care that is required and to pay attention to each individual's requirements. They will be pleased to provide any special assistance that you may need.

Patients with A-T will be accommodated in one of the side rooms, each of which has a roll-away bed for a family member or carer.

## Respiratory Infection, Inflammation, and Immunity (RIII) Service

This includes the adult Cystic Fibrosis unit, the Lung Defence and Immunology clinics and inpatient beds. The RIII attracts supra-regional and national referrals because of its expertise in the management of respiratory and immune problems. It has expertise in national clinic databases, whilst the Lung Defence and Immunology telephone support services deal with over 300 enquiries per quarter.

**Patients with A-T** will be seen in the RIII clinics for immunological assessment and to identify opportunities to support the immune system with the optimal use of standard vaccines, antibiotics, anti-virals, and immunoglobulin replacement as appropriate.

## The Multi-Disciplinary Team

Both the RSSC and RIII work in multi-disciplinary teams with excellent support from many disciplines. The programme includes assessment of swallowing by speech and language therapists by a Sips test and video fluoroscopy.

## Radiation Risks

A high-resolution CT chest scan and video fluoroscopy are standard investigations for assessment of chest problems, a major cause of ill health and death in A-T. We also know that A-T is a disorder associated with increased sensitivity to radiation at levels associated with radiotherapy, rather than diagnostic tests.

Discussions with A-T clinical experts at the international meeting in October 2004, at the Nottingham A-T Clinic, with Professor Malcolm Taylor, University of Birmingham and with the A-T Society have produced a consensus that a high-resolution CT chest scan and video fluoroscopy are low risk and essential to clinical assessment in A-T.

The CT scan and video fluoroscopy will be assessed by our radiologists who have considerable expertise in chest problems.

**The team are sensitive to concerns about radiation risks and are happy to discuss this. No examinations will be carried out without the patient's consent.**

## Referral

Adults (16 years plus) with A-T should be referred to the clinic by their local GP or hospital doctor. They can also be referred by Dr Mohnish Suri, Nottingham City Hospital if they have attended the A-T Clinic based there. Referrals should be addressed to:

Dr Andrew Exley  
Consultant Immunologist

Dr John Shneerson  
Consultant Chest Physician

Adult A-T Services  
Papworth Hospital  
Papworth Everard  
Cambridge  
CB23 3RE

The Papworth team will collate information from the referring doctor and the Nottingham A-T Clinic, including any previous immunological assessments.

**Any family seeking a referral should contact the A-T Society which will make the necessary arrangements.**

**The Society requests that any doctor referring a patient directly to Papworth Hospital also informs us so that we can assist with any necessary travel arrangements.**

## **The Ataxia-Telangiectasia Society** **Supporting the Family—Fighting the Disease**

**The Ataxia-Telangiectasia Society** is a national charity. Our objective is to alleviate the suffering and distress that A-T causes. We do this by:

- Supporting families
- Improving clinical management through two specialist clinics
- Providing information on the disease and raising awareness
- Funding research

**We rely totally upon voluntary donations to continue our work.**