9. Lung transplant assessment

Almost all assessments are now carried out at Great Ormond Street Hospital for Children and referrals should be made to Drs Helen Spencer or Paul Aurora. A referral proforma is available from Great Ormond Street Hospital (see below). An exception would occur in the case of an adolescent approaching transition to the adult service, in which case, the assessment should be done here, liaising with the adult team. Contact Dr Su Madge, Nurse Consultant, extension 4053 at Royal Brompton Hospital, for the booklet listing investigations. Once complete, return these to Dr Martin Carby or Dr Anna Reed, Consultants in Respiratory & Transplant Medicine, at Harefield Hospital.

Over the years, most transplants performed in CF children were heart / lung (HLT) with the CF patient's heart being used in a domino procedure for another patient. More recently, bilateral lung transplant is being done more often. Although living lobar transplants (a lobe each from two relatives, most commonly parents) have been performed in adults and some paediatric centres abroad, they are not yet performed in paediatric practice in the UK.

Consideration of a child for LT assessment should be based on the individual patient and is best performed in a multi-disciplinary fashion.

Criteria for Transplant Referral

- Significantly reduced lung function, usually with FEV_1 <30% predicted. May include rapidly declining FEV_1 even if still >30% predicted.
- Severely impaired quality of life.
- Oxygen-dependent (resting SpO_2 < 90%).
- Exacerbation of pulmonary disease requiring PICU/HDU stay.
- Pneumothorax in advanced disease especially if recurrent.
- Severe haemoptysis not controlled by embolisation.
- Child and family committed to the idea.

Traditionally, children fulfilling these criteria would be likely to have a median life expectancy of 2 years, but this may not be the case anymore.

Contra-indications

The following contra-indications differ between centres and may be subject to change over time with the availability of *e.g.* newer antibiotics and increasing surgical expertise. The decision will be influenced by the presence of multiple problems within an individual child.

1. Major

- Other organ failure (excluding hepatic when a lung/liver transplant could be considered).
- Untreated *Mycobacteria tuberculosis.*
- Invasive pulmonary aspergillosis.
- Malignancy in the last 2 years.
- Unstable critical clinical condition (*e.g.*, shock, mechanical ventilation or extra-corporeal membrane oxygenation).
- Infection with *Burkholderia cenocepacia* and *Mycobacterium abscessus* - all subspecies.
- Child does not want the procedure despite receiving and understanding information.

2. Relative

- Long term corticosteroids > 20mg/day.
- Non-pulmonary infections *e.g.* Hepatitis B or C, HIV.
- Previous thoracic surgery - pleurodesis will make the procedure more difficult and should be discussed with the surgical team.
- Multi-resistant organisms *e.g.* non-abscessus NTM, some genomovars of *B cepacia* complex, *MRSA*, pan-resistant *P aeruginosa*, treatment-resistant fungi.
- Severe osteoporosis.
- Some extreme psycho-social issues, for example, long standing and entrenched non adherence to treatments; lack of family support.

Transplantation is so familiar to many people now from TV, newspapers etc, most of which tend to be biased towards successful outcomes, that it is often perceived as a miracle cure. It is therefore important when discussing the issues with the family and child, that as well as the potential benefits, the following negative points should be addressed (these will be addressed at the assessment meetings, but should be raised early with families):

1. Acceptance onto the waiting list does not guarantee a transplant. Due to a shortage of donors about 25% of patients will die before organs become available. The time spent waiting for organs may be stressful (uncertainty, false alarms etc).

2. Lung transplantation is not a complete cure for CF, it is a palliative procedure. After the operation, invasive procedures including bronchoscopy and biopsies are required. In addition, unless complete eradication of reservoirs of infection has been successful (which almost never occurs due to chronic infection of sinuses), there is potential for bacterial infection of the transplanted lungs, which may make ongoing nebulised antibiotic therapy and physiotherapy necessary.

3. Transplantation has little impact on the non-pulmonary manifestations of the disease (i.e., enzyme replacement and other therapies need to be continued), although there may be nutritional benefits in the medium term. CF-related diabetes may worsen or develop.

4. Problems associated with transplantation include early rejection, severe sepsis related to immunosuppression and later development of obliterative bronchiolitis (OB). OB can eventually lead to severe respiratory impairment and is difficult to treat successfully.

UK Paediatric Lung and Heart-Lung Transplantation

Referral Proforma

STRICTLY CONFIDENTIAL

THIS FORM MAY BE USED TO REFER TO ANY OF THE UK CENTRES THAT PERFORM LUNG & HEART-LUNG TRANSPLANTATION. PLEASE RETURN THE FORM TO THE CENTRE OF YOUR CHOICE:

GREAT ORMOND STREET
Dr Paul Aurora and Dr Helen Spencer
Cardiothoracic Transplant Office
Great Ormond Street Hospital
Great Ormond Street
London
WC1N 3JH

Tel: 020 7813 8563
Fax: 020 7813 8440

NEWCASTLE
Dr Malcolm Brodlie
Cardiopulmonary Transplant Unit
Freeman Hospital
High Heaton
Newcastle upon Tyne
NE7 7DN

Office: 0191 223 1132
Fax: 0191 223 1439

GUIDANCE NOTES FOR
COMPLETION OF REFERRAL PROFORMA

This proforma has been designed to streamline the referral process for potential lung and heart-lung transplant recipients. As a result, potential transplant candidates can be identified more easily, be formally assessed more quickly and duplication of investigations will be avoided. The information required has been agreed by all UK lung transplant centres and this form can be used to refer to any UK centre.

Thank you for your co-operation.

KEY POINTS

Please complete all sections - any questions which are not applicable should be marked as N/A**.**

When specific results are not available but have been requested please mark as **awaited.**

Copies of Imaging (CT, coronary angiography, etc) should be sent on CD with this form

Copies of complete reports of investigations can be appended to this proforma, but the clinical summary should be completed by a member of the multidisciplinary team in the appropriate proforma section. Serial lung function tests are very helpful and should be included when available.

Any questions about this proforma or its use can be addressed by contacting the transplant co-ordinators at the hospital to which you intend to send the referral.

PERSONAL DETAILS

PATIENT NAME: …….

NHS Number:……

AGE: ……

DOB: ……

ELIGIBILITY FOR NHS CARE:……..

NEED FOR INTERPRETER: YES / NO LANGUAGE:……..

ADDRESS: ……

(Include Postcode) ……

……

TELEPHONE NUMBER ……MOBILE: ……..

REFERRING CONSULTANT:……..

REFERRING CENTRE:……

(Include Postcode) ……

……

TELEPHONE NUMBER ……FAX: ……..

PCT: ……..

GP NAME: ……

GP ADDRESS: ……

(Include Postcode) ……

……

GP TELEPHONE NUMBER: ……FAX: ……..

IS PATIENT AWARE OF REFERRAL FOR TRANSPLANT ASSESSMENT?

YES NO (please circle)

RESPIRATORY HISTORY

Primary Diagnosis: ……..

Secondary Diagnoses Respiratory…….

Non respiratory 1……

2…….

3…….

Respiratory Diagnoses made by: Clinical /CT chest/Histology/Genotype/ Sweat Test
Details…….

Any household members smoke? : YES NO (Please Circle)

Microbiology: Have these organisms ever been isolated?

Burkholderia cepacia *YES* *NO* *specimen*……date……

Pan-resistant Pseudomonas *YES* *NO* *specimen*……date……

MRSA *YES* *NO* *specimen*……date……

Mycobacteria (TB or atypicals) *YES* *NO* *specimen*……date……

Aspergillus *YES* *NO* *specimen*……date……

If YES, please give further details…….

……

Oxygen at home YES NO (Please Circle)

Amount …….L/min Average daily use …… hrs

Respiratory past history

Haemoptysis YES NO (Please Circle)

Details: ……..

Pneumothorax: YES NO (Please Circle)

Details: ……

Thoracic Surgery: YES NO (Please Circle)

Details: ……
Has the patient ever required ventilation? YES NO (Please Circle)

If yes NIV / formal ventilation in ITU (duration ……days)

Details:…….

Current Exercise Capacity

Exercise tolerance …… (distance)

Formal 6 minute walk test performed? YES NO (Please Circle)

If yes Max distance …… metres Lowest saturation……%

Performed on air / oxygen at …… litres per minute

Wheelchair YES NO

Progress pre- and post-diagnosis

Include details on rate of decline, life threatening exacerbations, frequency of IV antibiotics, etc

Is family aware of prognosis? YES / NO

Is patient aware of prognosis? YES / NO

PAST MEDICAL HISTORY

Current or previous :			Details:
Heart Disease	YES	NO	……
Renal Disease	YES	NO	……
Liver Disease	YES	NO	……
Diabetes	YES	NO	……
Malignancy	YES	NO	……
GI problems	YES	NO	……
Portacath	YES	NO	……
Gastrostomy	YES	NO	……

Current Medication

1…….	Dose	Frequency
2…….	Dose	Frequency
3…….	Dose	Frequency
4…….	Dose	Frequency
6…….	Dose	Frequency
7…….	Dose	Frequency
8…….	Dose	Frequency
9…….	Dose	Frequency
10……..	Dose	Frequency

ALLERGIES: YES NO (Please Circle)

1……

2……

Oral Corticosteroids? YES NO (Please Circle)

Date commenced

Max dose Current dose Date stopped

Response…….

Family and Social History

Adherence Good YES NO (Please Circle)

Attendance Record Good YES NO (Please Circle)

Family support available:……

Social Services input: YES NO

Details……..

School details:…….

School attendance:……..

Siblings?.....

Relevant Family Medical or Social History:……..

……
……

……
……

……
……

……
……

……
……

Psychological assessment

Current or Previous History of:

Depression: YES NO
Panic attacks: YES NO
Anxiety: YES NO
Needle phobia: YES NO
Other psychological concerns?: YES NO

Details ……...

……

……..

……..

……..

……..

CLINICAL INVESTIGATIONS

Weight…….kgs Height…….m BMI……..

ECG Date performed:

Result……. ……..…

Echocardiogram Date performed:

Result……...

Chest x-ray Last performed:

Result……...…

HRCT Thorax Date performed

Result……...…

……..

Arterial/Capillary/Venous (please circle) Blood Gas (ON AIR)

pH …….. pO2 …….. pCO2 …….. BXS …….. HCO3 …….. Sats …….

Others (**if available**)

Bone Densitometry Spine Z score = ……..… Femur Z score = ……..…

Abdominal ultrasound ……..

Coronary angiography ……..

Right heart catheter ……..…

GORD Testing ……

Glomerular Filtration Rate …..……

Respiratory Function Tests **(attach trend values if possible)**

Date	…….		…….	
	Value	%	Value	%
FEV1	…….	…….	…….	…….
FVC	…….	…….	…….	…….
FEV1/FVC	…….	…….	…….	…….
TLC	…….	…….	…….	…….
FRC	…….	…….	…….	…….
RV	…….	…….	…….	…….
TLCO	…….	…….	…….	…….
KCO	…….	…….	…….	…….

Haematology	
Date:	
Na	
K	
Urea	
Creatinine	
eGFR	
Bilirubin	
ALT	
ALP	
GGT	
Glucose (fasting)	
Chol (fasting)	
Trig (fasting)	
Total Calcium	
CRP	

Biochemistry	
Date:	
Hb	
WCC	
Platelets	
PT	
APTT	
Fibrinogen	
ESR	

Virology	
Date:	
HIV	
CMV	
Hepatitis B	
Hepatitis C	
Immunology	
IgE	

Additional Microbiology	
	Date & Details
MRSA screen	
Asp. precipitins	
Asp. culture	

Blood group (if known) ……..

Anti crossmatch antibodies (if known) YES NO

Details ……..

ANY OTHER COMMENTS

Signed……. NAME:……..

POSITION:……. DATE:……