

CF Guidelines - Transplantation

Referral for Lung Transplantation:

Lung transplantation is an option for some patients with end stage lung disease. However, not all patients are eligible. Other, fully informed patients do not feel that it is the right choice for them. Their decision should be respected.

Timing Referral:

Data from the USA (Nathan, 2005) suggests that average wait between listing for lung transplantation and operation is 2 years. Referral too late may mean that patients die before an organ becomes available. In the UK 50-60% of suitable patients die before a donor organ becomes available. Conversely, referral too early may result in transplanted patients having a worse outcome than those who have not had surgery. Liou et al, 2007, suggest that only children with a predicted 5 year survival of 0-30% have a survival benefit from transplantation. Traditionally patients with CF are referred for transplantation when their FEV1 falls consistently below 30% predicted. However, on an individual basis, this is a relatively poor predictor of death within 2 years.

Although there are no consistent data, additional factors which may be considered:

- Rate of decline in lung function.
- Increasing frequency of admissions.
- Deteriorating quality of life.
- Declining body mass index.
- Pneumothorax.
- Female gender.

Making a Referral:

- For reasons of geography, all referrals for lung transplantation in the South West are made to Brompton/Harefield. Their guidance on making a referral is reproduced in full towards the end of this document. However, other UK centres may take differing views of the impact of medical co-morbidities on eligibility for transplantation. So, if a patient is deemed ineligible by Brompton/Harefield, the views of Papworth and Newcastle may be sought, although acceptance by these centres is likely to require the patient to move and live within a appropriate travelling distance (usually within 4 hours).
- Generally steroid usage should be limited to 10mg od Prednisolone or less.
- Generally patients must be free from substance addiction for a minimum of 6 months.
- Generally patients should have been free?

The Operation:

In the UK the vast majority of patients with CF receive a bilateral sequential lung transplant. Heart-lung transplantations are now much less common. Living donor transplants have been performed (2 donors each donate a lower lobe). But to date, survival outcomes have been worse than for cadaveric transplantations, although the subsequent incidence of post-transplantation obliterative bronchiolitis may be reduced.

Outcome and complications:

Newcastle (Meachery et al, 2008) recently reported values of 82% survival at 1 year, 70% at 3 years, 62% at 5 years and 51% at 10 years. Bronchiolitis obliterans syndrome (BOS) and sepsis were the common causes of death. Freedom from BOS

was 74% at 5 years and 38% at 10 years. Infection with *Burkholderia cenocepacia* (genomovar 3) was associated with a worse outcome. Some UK centres will no longer consider patients infected with this pathogen.

References:

- 1, Liou TG et al. N Engl J Med 2007;357:2143-52.
- 2, Meachery G et al. Thorax. 2008; 63:725-31.
- 3, Nathan SD. Chest 2005; 127:1006-16.

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