Lung transplantation for cystic fibrosis: satisfactory results in specialized centres

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Cystic fibrosis (CF) is a formidable genetic disease. Respiratory physiotherapy, care and management in specialized centres have considerably lengthened life expectancy.

The advances made in lung transplantation for CF can clearly be seen in the International Registry. It is particularly true for highly specialized teams like that of Zurich. CF is their primary indication for transplantation with excellent results at 1 and 5 years presented in this issue of *EJCTS* [1]. Improved results have also been noted for our team [2], which has carried out 185 transplants for CF to date.

Management by teams of specialized paediatricians and pulmonologists makes it possible to carry out transplantation as late as possible, but not too late.

Modifications to the graft allocation systems in the United States and several European countries allow transplantation of the most seriously ill patients [3]. Use of grafts with extended donor criteria, and evaluation of very marginal grafts with an *ex vivo* reperfusion machine, should limit preoperative deaths by increasing the pool of grafts available.

Some problems are specific to transplantation for CF [4]. The patients are usually underweight which increases the risk of postoperative mortality [5]. Therefore, a programme of hyper-nutrition and physiotherapy should be set up during the waiting time.

Some patients have a Burkholderia cepacia complex (BCC) chronic lung infection. A high mortality rate after transplantation is associated with Burkholderia cenocepacia infection (and these patients are excluded from transplantation in many teams) but not with non-cenocepacia BCC species [6]. Infection with *Burkholderia gladioli* increases morbidity.

In case of infection with *Mycobacterium abscessus*, a multi-resistant mycobacteria, pre-transplant eradication therapy should be attempted [7]. Long-term treatment with multiple drugs may be complicated in the CF patient with severe disease.

In some severe patients listed for transplantation, mechanical ventilation and extra corporal membrane oxygenation must be instituted. In these cases, a rise in postoperative mortality has been reported in the International Registry [8]. However, if transplantation is carried out in the case of pulmonary failure alone,
the results are acceptable (no increased postoperative mortality in our experience). Several countries including Switzerland have set up a system of lung allocation priority for this type of highly urgent transplant candidate. Four patients were transplanted in these conditions in the series reported by the Zurich team.

Some patients have liver disease associated with portal hypertension. A combined lung and liver transplantation is necessary in some cases [9] but can be avoided when the liver disease is not too severe [10].

CF patients are often small, and size reduction techniques and lobar transplantation have progressively been adopted by all the specialized teams.

Cardio-pulmonary bypass (CPB) has not been routinely used in the Zurich experience, but only according to the haemodynamic and gas exchange with good results, and so the debate on the routine use of CPB is ongoing.

A zero complication rate for bronchial anastomoses is reported in these series. All over the world, bronchial anastomoses are performed nowadays very near to the upper lobe bronchus, resulting in very few bronchial anastomosis dehiscences, visible on computed tomography or fibrescopy, which disappear spontaneously. However, bronchomalacia and bronchial stenoses sometimes occur at the level of the bronchus intermedius or lobar bronchi, distant from the bronchial anastomosis, and require bronchial dilation and stenting. None of those complications are reported in the series presented here.

The Zurich team is to be congratulated for these excellent results which mean that lung transplantation has become for these young patients the hope for a better life rather than something to be feared.

REFERENCES