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Outcome after Liver Transplantation for Cystic Fibrosis

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Introduction

Improvements in pulmonary care and other refinements in management allow more patients with cystic fibrosis (CF) to survive long enough for liver disease to become a limiting factor in their life expectancy (1, 2, 3). Since liver transplantation is considered the treatment of choice for patients with end-stage liver disease, it is predictable that more patients with CF and liver failure will be referred for transplantation. The results after liver transplantation in 14 patients with CF are reviewed in this report.

Clinical Material

Between January, 1981 through May, 1990 3,019 liver transplantations were performed at the University of Pittsburgh. Eighteen of these transplants were performed in 14 patients (6 adults and 8 children) with CF who had developed end stage liver disease. The first nine patients, previously reported in 1989 (3), were done under cyclosporine, prednisone and azathioprine inmmunosupression. The last 5 patients were done with the new immunosuppressive drug FK506 and low dose prednisone. The patient demographics, including pre- and postoperative pulmonary function tests and current patient status, are summarized in table 1 below. One patient, who received a a double organ transplant (liver and pancreas), is not included in the present series.

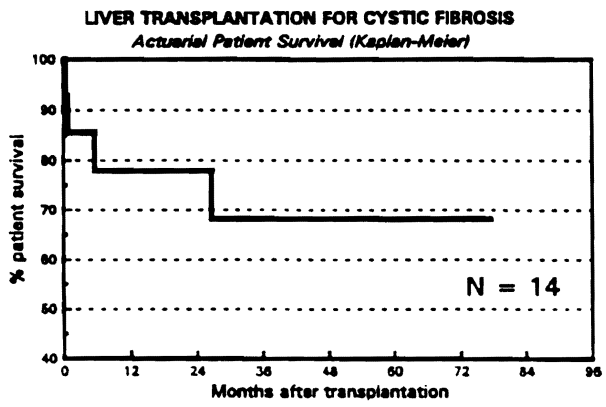
Table 1. Patient Demographics, Pulmonary Function, and Current Status

Patient	Age/Sex	FVC/FEV_1		Current Status
		Preoperative	Postoperative	
1	23y/M	72/81	109/119	Alive at 7 1/2 yr, working full time
2	30y/F	62/50	59/43	Died at 27 months
3	24y/M	105/114	110/105	Alive at 65 months, working full time
4	15m/F			Alive at 60 months, attending school
5	3y/M			Alive at 43 months, attending school
6	21y/M	77/74		Died at 2 weeks
7	8y/M	50/38	109/106	Alive at 36 months, attending school
8	13y/F	81/68	114/101	Alive at 31 months, attending school
9	18y/F	61/50	81/70	Alive at 31 months, attending college
10	31y/F	77/66		Died at 5 1/2 months
11	6y/M	114/94		Died at 2 weeks
12	14y/M	86/70	94/83	Alive at 10 months, attending school
13	12y/M	90/71		Alive at 10 months, attending school
14	13y/M	53/51	58/45	Alive at 2 months, recuperating

Results

Ten of the 14 recipients are well with normal liver function tests 2 months to 7.5 years after liver transplantation. Two patients died in the early postoperative period of sepsis and multiple organ failure following retransplantation, one with hepatic artery thrombosis (patient #6) and the other (patient #11) with severe allograft rejection. A third gravely ill patient (patient #10) with advanced liver disease and severe exocrine and endocrine pancreatic

insufficiency, died with a perfectly functioning graft 6 months after transplantation. The patient, who required a femoral embolectomy on the second postoperative day, remained ventilatory dependent thereafter and died of respiratory failure. A fourth recipient (patient #2) died 27 months after transplantation from an unrecognized closed-loop obstruction and gangrenous bowel. This patient also had Friederich' s ataxia and mental retardation.



Actuarial patient survival (Kaplan-Meier) for the 14 patients in this series is shown in the figure to the left and is 77.9% at one year and 68.2% at 5 years. Pulmonary function tests were done in eight patients before and after transplantation (see table 1). Forced vital capacity (FVC) and first second forced expiratory volume (FEV_1) were improved in patients 1, 7, 8, 9,and 12, and essentially unchanged in patients 2, 3, and 14.

Discussion

The long-term patient survival rate of nearly 70% in this series of patients demonstrates that results for patients with cystic fibrosis are comparable to those reported for other established indications for liver transplantation (4). Nevertheless, due to the multisystemic nature of CF, these patients are more likely to develop complications in the early postoperative period. Therefore, in order to improve results in this group of patients, the following points should be stressed: 1) the need for meticulous pulmonary toilet in every patient, and for early tracheostomy in the patients who require reintubation or reoperation; 2) aggressive bowel care and early introduction of parenteral nutrition; and 3) when using cyclosporine, which is poorly absorbed in this group of patients, close monitoring of cyclosporine blood level and dosage is recommended. Our experience with liver transplantation for CF using FK 506 is still small, but encouraging.

Mild to moderate pulmonary disease is not a contraindication to liver transplantation in patients with CF. In fact, some improvement in pulmonary function was noted in 5 of the 8 patients who were tested before and after transplantation. The reason is not clear yet, although there are at least two explanations: 1) improved ventilatory mechanics secondary to disappearance of ascites, rebuilding of thoracic muscle mass, and relief of fatigue; and 2) modulation of the endobronchial inflammation by immunosuppressive agents.

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