



Empirical Articles

Quality of Life Following Liver Transplantation in Patients With Familial Amyloid Neuropathy

A Prospective Controlled Study

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Abstract

Aim: The present study aimed to evaluate the change in quality of life 12 months following liver transplantation in patients with Familial Amyloid Polyneuropathy (FAP).

Method: In this study 150 transplant candidates, attending the outpatient clinic of a Liver Transplantation Centre in Lisbon, were assessed between March 1, 2006 and December 1, 2007. From these, 84 were transplanted, and 62 finished the study; 20 with FAP and 42 with Liver Disease (LD). These patients were assessed before, and 12 months after, transplantation. The patients that remained waiting for transplantation originated the control group. First, transplanted (study group) and non-transplanted (control group) patients were compared regardless of their diagnosis, and then only FAP patients were compared between both groups.

Results: 12 months after transplantation the score on the Quality of Life's Physical and Mental Component of the SF-36 was significantly higher in transplanted versus non-transplanted patients (concerning the whole group FAP and LD patients). However, significant differences were only found for the Quality of Life's Physical Component subscale between both FAP groups (study and control group).

Conclusion: In sum, liver transplantation does not have a significant impact in FAP patients' Mental Quality of Life score. One possible reason to this is the fail in acquiring adaptive coping strategies after transplantation.

Keywords: liver transplantation, familial amyloid polyneuropathy, quality of life, mental health, coping

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Introduction

Familial amyloid polyneuropathy (FAP) is an autosomal dominant multisystemic fatal disorder characterized by a progressive peripheral and autonomic neuropathy with neural and systemic amyloid deposits (Monteiro, Freire, & Barroso, 2004). The amyloid protein in type 1 FAP of Portuguese, Swedish and Japanese origin, is the variant of transthyretin (TTR), in which methionine a substitute for valine at position 30 is (TTR Met). Most of this TTR Met 30 is produced by the liver. The most consensual way to treat FAP is liver transplantation in the initial stage of the disease, to prevent neurological deterioration.

Patients with FAP are almost asymptomatic when they are transplanted in contrast to other liver transplant candidates that usually have a disabling chronic liver disease (Monteiro et al., 2004). It has been demonstrated that quality of life (QoL) can be improved after transplantation in patients with liver disease (Monteiro et al., 2004; Price et al., 1995; Riether, Smith, Lewison, Cotsonis, & Epstein, 1992; Telles-Correia, Barbosa, Mega, & Monteiro, 2008). To our best knowledge, no other study was conducted besides by our group that evaluates QoL improvement in FAP patients. Nevertheless, this was done by comparing QoL 6 months after transplantation in patients with FAP and those with chronic liver disease (Telles-Correia, Cortez-Pinto, Barbosa, Mega, & Monteiro, 2009).

The aim of the present prospective study was to evaluate the change in quality of life 12 months after liver transplantation in patients with FAP. This was accomplished using a group of non-transplanted FAP patients as control group.

Method

Participants

A group of 150 transplant candidates, attending the outpatient clinic of Liver Transplantation Centre in Lisbon, were studied between March 1, 2006 and December 1, 2007. From these, 84 were transplanted.

These patients were assessed twice (the first time before transplantation and the second time 12 months after transplantation). Within the transplanted group, 11 patients died, three were retransplanted (thus, excluded from the study), and eight withdrew from the study. A total of 62 transplanted patients finished the study (study group). The patients that remained waiting for transplantation originated the control group.

Written informed consent was obtained from all participants, and the study protocol was approved by the Curry Cabral Hospital's Ethical Committee.

Regarding medical diagnosis, within the study group, 20 patients had FAP and 42 had chronic liver disease (CLD). Among the patients with liver disease, 16 had Alcohol-related Liver Disease (ALD), six Hepatitis C and Associated Cirrhosis (HCAC), one Hepatocellular Carcinoma (HCC), one Primary Biliary Cirrhosis (PBC), one Progressive Familial Intrahepatic Cholestasis (PIC), one Sclerosing Cholangitis, one Cryptogenic Cirrhosis (CC), and the remaining had Mixed Diagnoses (11 ALD + HCC; seven HCAC + HCC; three HCAC + HCC + ALD; one HCC + Virus B Liver Disease; one Hemochromatosis + HCC), three unknown cause. Within the control group 15 patients had FAP and 25 had CLD. Among the patients with CLD, 11 had ALD, two HCAC, two PBC, one PIC, one CC, and the others had Mixed Diagnosis (six ALD + HCC; seven HCAC + B Hepatitis).

Within the study group, in the CLD subgroup, 18 patients were classified Child-Pugh class A (see Medical evaluation chapter for the explanation of this classification), 22 class B and six class C. In the FAP subgroup ten patients were classified as Level 1 FAP, eight Level 2 and two Level 3. Within the control group, in the CLD subgroup, ten patients were classified Child-Pugh class A, 11 class B and four class C. In the FAP subgroup, Nine patients were classified as Level 1 FAP, five Level 2 and one Level 3.

In the study group 40 patients were male, and the mean age was 45.40 ($SD = 12.22$). In the control group, 24 patients were male and the mean age was 47.20 ($SD = 13.33$). No significant differences regarding demographic and medical data were found between both groups.

Instruments

Quality of Life — We used the MOS 36-Item Short Form Survey Instrument (SF-36; Ware, 1993). Portuguese validated version (Pais-Ribeiro, 1993), a self-rating questionnaire developed by the Medical Outcome Trust (Ware, 1993), to assess quality of life. The SF-36 has been widely used under a range of different medical conditions and shown to have adequate reliability and validity. The first four subscales refer to physical aspects, and the last four scales, mental aspects: physical functioning, physical role Limitation, bodily pain, general health, vitality, social functioning, emotional role limitation and emotional well-being. Physical aspects mostly refer to physical capability to perform normal daily activities. The mental dimension mostly refers to social aspects of life the degree to which illness interferes with emotional well-being, and social roles. Total values were computed for physical and mental components of health-related quality of life by averaging the eight weighted subscales.

Medical Evaluation — The diagnosis of chronic liver disease (CLD) was established by a hepatologist. Child-Pugh classification was used to evaluate liver disease severity. It is based on a patient's bilirubin and albumin serum levels, the international normalized ratio (INR) (a measure of coagulation) and the presence or absence of ascites and encephalopathy. Patients belonging to class A are the ones with best prognosis and the ones belonging to class C have the worst prognosis.

The diagnosis of FAP was established by a neurologist. To evaluate the severity of FAP the (REF) (Sales-Luís, Galvão, Sousa, Alves, & Serrão, 1990) Portuguese classification was used. This scale considers six levels of severity: Level 0 – asymptomatic; Level 1 – sensitive and/or dysautonomic symptoms without neurological signs; Level 2 – sensitive and/or dysautonomic symptoms + neurological signs (sensitive); Level 3 – sensitive and/or dysautonomic symptoms + neurological signs (sensitive-motor) in lower limbs with independent walking; Level 4 – neurological signs in lower and upper limbs (sensitive or motor) walking without help; Level 5 – neurological signs (sensitive or motor) in lower and upper limbs, in wheel chair; Level 6 – confined to bed.

Procedure

Patients from the study group (transplanted patients) were assessed twice: the first time before transplantation and the second time 12 months after transplantation. The ones that remained waiting for transplantation originated the control group.

Firstly, transplanted (study group) and non-transplanted (control group) patients were compared regardless of their diagnosis, and then only FAP patients were compared between both groups.

Statistical Analysis

Statistical analyses were performed with the SPSS 13.0 for Windows software package. Descriptive data were presented in absolute frequencies, percentages, mean values, standard deviations, and modes. QL differences between groups were assessed by means of the Mann Whitney test ($p < .05$ was considered statistically significant).

Results

Quality of Life Differences

First, we looked for the differences between the study group and the control group within the whole sample. We found that before transplantation there were no differences between the groups. Quality of Life's Physical and

Mental Component was significantly higher in the study group (Table 1) 12 months after transplantation. Regarding FAP patients, before transplantation there were no differences between the study group and the control group. After transplantation we still did not find significant differences for Quality of Life's Mental Component, though we did for Quality of Life's Physical Component (Table 2).

Table 1

QoL in the Total Sample 12 Months After Transplantation – Differences Between the Study Group (FAP + LD) and the Control Group (FAP + LD)

QoL	Study Group (FAP+LD)	Control Group (FAP+LD)	<i>U</i>	<i>W</i>	<i>p</i>
Physical component	68.29	43.15	359.00	887.00	.001
Mental component	70.12	55.63	361.00	922.00	.008

Table 2

QoL in the FAP Subgroup 12 Months After Transplantation – Differences Between the Study Group and the Control Group

QoL	Study Group (FAP)	Control Group (FAP)	<i>U</i>	<i>W</i>	<i>p</i>
Physical component	65.25	43.00	110.00	320.00	.010
Mental component	68.50	61.00	430.00	720.00	.800

Discussion

Several authors found an improvement in mental and physical quality of life components after liver transplantation (Price et al., 1995; Riether et al., 1992). However, most of them evaluated decompensated liver disease patients, and not special groups, such as FAP patients. To our knowledge, this is the first study evaluating mental and physical quality of life in FAP patients 12 months after liver transplantation, using non-transplanted FAP patients as a control group. FAP patients have a genetic disease, invariably fatal without treatment, and are now being transplanted at a very early stage, with a paucity of symptoms, and therefore a very special group.

Telles-Correia and colleagues (2008) found that there was a high prevalence of psychiatric diagnoses (mostly depression and anxiety) in familial amyloid polyneuropathy patients waiting for transplantation and that these patients had a different personality profile comparing to other liver transplant candidates. This might be due to the emotional stress of being exposed to a chronic sensation of being carrier of a fatal genetic disease (Telles-Correia et al., 2008)

The fact that liver transplantation had a less beneficial impact in the mental quality of life of FAP patients is difficult to explain. One possible reason to this is the failure in acquiring adaptive coping strategies after transplantation. Telles-Correia, found that these patients could develop unadaptive coping styles after transplantation because of a poor familiarity with acute medical symptoms and due to unrealistic expectations towards liver transplantation (Telles-Correia, 2013). According to Moos and Schaefer's (1993) model, coping strategies can predict mental health and quality of life (Telles-Correia, 2013), and this might be an explanation for our conclusions, concerning FAP patients' quality of life after liver transplantation. These hypotheses must be confirmed by further studies.

Based on our findings, special groups such as FAP patients should be followed up with special attention since they may be more vulnerable in terms of mental health and quality of life.

One of the main limitations of the present study is the small number of FAP patients included, which relates to the difficulty of conducting a prospective study in such a rare disease. More studies in this area are needed to confirm our findings and further explore our knowledge about FAP patients' psychological profile, in order to design specific strategies that can improve their quality of life after liver transplantation.

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Competing Interests

The authors have declared that no competing interests exist.

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Notes

All authors have contributed to: providing conception and design; data acquisition; data analysis and interpretation; drafting the article; revising it critically for important intellectual content; contributing to statistical analysis.

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