ORIGINAL ARTICLE

Quality of life and illness perception in primary biliary cirrhosis: A controlled cross-sectional study

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Summary

Objective: The aim of this study was to understand better the quality of life (QOL) and illness perception in women with primary biliary cirrhosis (PBC) through a comparison with women having diabetes.

Methods: One hundred and ninety-four women took part in this study: 130 with PBC, 64 with type 2 diabetes. They were administered the SF-12 to measure QOL and the Brief Illness Perception Questionnaire to assess representations of their illness. Analysis of covariance with bootstrapping was used to compare QOL and illness perception scores by controlling age and mean disease duration.

Results: Physical QOL was significantly worse for women with PBC than for women with diabetes. Women with PBC felt their disease would last longer and reported more symptoms and concerns related to their disease than women with diabetes. Significant differences

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were also observed for causes: women with PBC mainly reported autoimmune, emotional, unknown/unlucky and medical causes whereas women with diabetes reported mostly lifestyle and hereditary causes. Marginally significant differences were observed regarding consequences on daily life, feeling of control over the disease and emotional responses, which were shown to be worse in PBC. Mental QOL, treatment control and overall understanding of the disease was similar in both groups.

Conclusions: This study shows that women with PBC have a worse QOL and somewhat different illness perception than women with diabetes. Further research could help understand PBC specificities better in order to improve patient care, especially if factors such as fatigue or rarity of the disease explain these results.

Introduction

Primary biliary cirrhosis (PBC) is a chronic cholestatic liver disease of unknown etiology whose prevalence ranges from 100 to 400 per million people [1]. It occurs more in women (90%) and is most often seen in middle age. PBC is slowly progressive and can result in cirrhosis and liver failure. Initially asymptomatic, patients will often develop pruritus and fatigue over the course of the illness [2]. Quality of life (QOL) is altered in patients with PBC compared to control groups from the general population matched for sex and age [3,4]. Fatigue is the symptom that seems to have the greatest impact on QOL in this population [4,5]. Altered QOL is not specific to PBC and is observed in many chronic diseases, such as diabetes, chronic kidney disease and heart failure [6—8].

According to Leventhal's Self-Regulation Model [9], when people are facing an illness, they create their own representations in order to make sense of the problem and to cope with it. Illness perception is composed of both cognitive and emotional representations. Five main components have been identified in cognitive representation:

- *identity*, which is the label the person uses to describe the illness and the symptoms they view as being part of the disease;
- *consequences*, which are the expected effects and outcomes of the illness;
- *causes*, which are the personal ideas about the causes of the illness;
- *timeline*, which is the length of time the patient believes the illness will last;
- *cure/control*, which is the extent to which the patient believes that he/she can recover from or control the illness.

Emotional representation includes negative reactions such as fear, anger and distress. Studies among patients with various chronic diseases have shown that illness perceptions are associated with different outcomes such as QOL. For example, in patients with type 2 diabetes, negative beliefs about illness are associated with low physical and mental functioning [10]. To our knowledge, illness perception has never been studied in PBC.

In order to understand better the specificity of PBC, and particularly the experience of living with a rare disease, the aim of the present study was to assess QOL and illness perception in patients with PBC through a comparison with patients having a common chronic disease. Diabetes is one of the most widespread chronic diseases in the population with a worldwide prevalence of around 8.3% in adults in 2013 [11]. People with type 2 diabetes often have no symptoms at first and may have none for many years. Early symptoms may include fatigue, hunger and increased thirst. As mortality and morbidity is higher in diabetes than in PBC [12,13], we hypothesized that diabetes patients would have a lower QOL than PBC patients. Moreover, we hypothesized that illness perception would be different in PBC compared to diabetes because of the stigma of living with a rare organ-specific disorder [14]. As PBC occurs more in women (90%), we decided to focus our study on them.

Materials and methods

Participants and data collection

Sample 1: PBC patients
PBC patients were recruited through the Hepatology Department of Saint-Antoine Hospital (Paris, France) and through a specific French patient association dedicated to the fight against inflammatory biliary diseases (''Association pour la Lutte contre les Maladies Biliaires Inflammatoires'', ALBI), specifically PBC, primary sclerosing cholangitis and autoimmune hepatitis. At the Saint-Antoine Hospital, patients with PBC were identified by physicians. The study was presented during a medical consultation and questionnaires were given with a pre-paid return envelope. For the association, an email was sent to all members to present the study and invite them to take part through an online survey. The distribution and collection of the questionnaires were carried out between March and September 2012.

One hundred and fifty-seven patients with PBC took part in the study. For the purpose of the present research, PBC patients with autoimmune hepatitis overlap syndrome were

excluded, as were men, questionnaires with missing data, and patients with a history of liver transplantation. A total of one hundred and thirty women remained. Most women were recruited through patient association (91.5%).

Sample 2: diabetic patients

As part of a larger study on the ability of general practitioners to detect core symptoms in patients, patients treated for diabetes type 2 were recruited by their general practitioners in several towns all over France. Inclusion criteria were: being treated for diabetes for at least one year, aged between 40 and 70 years old and without any comorbidity except micro- and macro-angiopathy. The administration and collection of the questionnaires were carried out between September 2011 and March 2012. Two hundred and forty-nine patients took part in the study. For the purpose of the present research, men and questionnaires with missing data were excluded. A total of sixty-four women remained.

Questionnaires

The *Short Form 12* (SF-12) is a general QOL instrument derived from the SF-36 [15]. It consists of 12 items that measure two dimensions: physical QOL and mental QOL. Scores range from 0 to 100 with a higher score indicating a greater QOL. The French adaptation has satisfactory psychometric properties [16].

The *Brief Illness Perception Questionnaire* (Brief-IPQ) [17] is a short version of the Illness Perception Questionnaire [18]. It consists of 9 items that measure:

- *consequences* (How much does your illness affect your life?);
- *timeline* (How long do you think your illness will continue?);
- *personal control* (How much control do you feel you have over your illness?);
- *treatment control* (How much do you think your treatment can help your illness?);
- *identity* (How much do you experience symptoms of your illness?);
- *concern* (How concerned are you about your illness?);
- *understanding* (How well do you feel you understand your illness?);
- *emotional response* (How much does your illness affect you emotionally?) (e.g. Does it make you angry, scared, upset or depressed?);
- *causes* (List in rank-order the three most important factors that you believe caused your illness).

Except for the *causes* item, which is an open question, the 8 other items have a numerical response scale ranging from 0 to 10. The scale does not have any cut-off. In order to rank patient scores for each numerical item, the following criteria were used: scores lower than 2 were considered ''weak'', scores between 2 and 4 were considered ''quite weak'', scores between 4 and 6 were considered ''moderate'', scores between 6 and 8 were considered ''quite high'' and scores over 8 were considered ''high''. The Brief-IPQ has been adapted in French by Broadbent and

is available on the Illness Perception Questionnaire website: http://www.uib.no/ipq/.

Statistical analysis

In order to compare sociodemographic and clinical characteristics of PBC and diabetes women, Chi^2 tests and analyses of variance were used. Our two samples were not matched regarding size, age and disease duration. Therefore, analysis of covariance with bootstrapping was used to compare quality of life and illness perception scores, by controlling age and mean disease duration. The bootstrap bias-corrected and accelerated interval was used as it is generally considered superior to other methods [19]. A total of 1000 replicates was generated from the original dataset in order to compute 95% confidence intervals.

Regarding the illness perception item measuring *causes*, a content analysis was first carried out to categorize answers and then a Chi^2 test to compare both diseases. Statistical analyses were performed using SPSS, version 17 (IBM, NY, USA). *P* values equal to or less than 0.05 were considered statistically significant. Continuous variables were expressed as mean ± standard deviation and categorical variables as a percentage.

Results

Patient characteristics

The mean age was 57.2 ± 11.7 years, with PBC women being significantly younger than diabetes women (54.5 vs. 62.8; *P* < 0.001) (Table 1). In both diseases, most women were in a couple (68.8%) and had at least one child (85.5%). Regarding occupational status, 48.1% of women with PBC and 35.9% with diabetes were employed. This difference was not statistically significant (*P* = 0.108).

The mean duration of illness was 8.7 ± 6.9 years, with PBC women having a shorter mean duration than women with diabetes (8 vs. 10.1; *P* < 0.05). Most women with PBC had an early stage disease (I, 24.1%; II, 12.3%; III, 8.7%; IV, 5.1%; unspecified, 25.2%) when the diagnosis was made. All were treated with ursodeoxycholic acid (13−15 mg/kg/d). A minority of women with diabetes had micro- or macro-angiopathy (15.7%) and were insulin-dependent (6.3%).

Quality of life

Physical QOL was significantly worse for women with PBC than for women with diabetes (39.8 vs. 44.7; *P* = 0.001; Tables 1 and 2). No significant difference was observed for mental QOL (*P* = 0.408).

Illness perception

Consequences on women's lives were perceived as moderate (5.1 out of 10 for PBC and 4 for diabetes) (Tables 2 and 3). Women with PBC perceived more consequences on their life than women with diabetes, but the difference was marginally significant (*P* = 0.064).

Table 1 Sociodemographic and clinical characteristics of participants by disease.

	Total	PBC	Diabetes	Chi^2/F	P value
n	194	130	64		
Mean age (SD)	57.2 (11.7)	54.5 (11.4)	62.8 (10.4)	24.49	0.000
Marital and family status					
Living in a couple (%)	68.8	66.4	68.8	0.107	0.744
Has at least one child	85.5	85.3	85.9	0.015	0.902
Occupational status (% employed)	44.1	48.1	35.9	2.576	0.108
Mean disease duration (SD)	8.7 (6.9)	8.0 (6.7)	10.1 (7.0)	4.235	0.041

Timeline was high (9.9 for PBC and 9.1 for diabetes), which means that women perceived their illness as continuing forever. Women with PBC reported their disease as being significantly longer than women with diabetes (*P* = 0.006).

Personal control was moderate (5.3 for PBC and 6.1 for diabetes). Women with PBC reported a lower *personal control* than women with diabetes but the difference was marginally significant (*P* = 0.060).

Treatment control was perceived as high (8.2 for PBC and 8.5 for diabetes), which means that women felt their treatment was helping. No significant difference was observed (*P* = 0.234).

Identity was quite weak (4.3 for PBC and 2.8 for diabetes) and women with PBC experienced significantly more symptoms of their disease compared to those with diabetes (*P* = 0.005).

Concern was quite high (8 for PBC and 6.5 for diabetes). Women with PBC were significantly more concerned about their disease compared to women with diabetes (*P* = 0.022).

Understanding was also quite high in both diseases (7.1 for PBC and 7.4 for diabetes). No significant difference was observed (*P* = 0.207).

Emotional response was moderate (6.0 for PBC and 4.5 for diabetes). Women with PBC were more affected emotionally by their illness than women with diabetes but the difference was marginally significant (*P* = 0.077).

Regarding *causes*, only the most important cause was analyzed as many subjects indicated only one cause (Table 3). Answers were categorized into six groups:

- *genetics*, which grouped the answers ''genetic'', ''hereditary'' and ''autoimmune'';
- *lifestyle*, which grouped ''low physical activity'', ''food'', ''overweight'' and ''tobacco'';
- *emotion*, which grouped ''shock'', ''event'' and ''stress'';
- *medical*, which grouped ''medication'', ''vaccine'', ''transfusion'' and ''pregnancy'';
- *unknown or unlucky*;
- *age*.

Significant differences were observed (*P* < 0.001). Women with PBC believed that their illness was mostly due to *emotion* (20.6%) or *genetics* (20.1%), with *autoimmune* being the most reported word. *Unknown or unlucky* (12.2%) and *medical* (10.6%) were also important causes whereas *age* was little reported (3.7%) and *lifestyle* was almost absent (0.5%). Women with diabetes believed their illness was mostly due

to *lifestyle* (12.7%) or *genetics* (11.6%). *Age* (3.7%), *emotion* (2.1%) and *medical* (1.6%) were a little reported while *unknown or unlucky* was almost absent (0.5%).

In addition, an analysis of PBC patients without cirrhosis (stage disease IV) was performed. Results did not change (Tables 2 and 3).

Discussion

This study shows that women with PBC have a worse QOL and illness perception than women with diabetes. Previous studies have shown that patients with PBC have a worse QOL than control groups from the general population, thus this study adds that, compared to a well-known slowly progressive chronic disease such as diabetes, women with PBC have an even worse physical QOL. It is also the first study to investigate illness perception in PBC. In particular, we observed that women with PBC reported being more concerned by their illness and experiencing more symptoms of it.

The results showed that physical QOL in women with PBC was worse than in those with diabetes, whereas mental QOL was similar. This is surprising as diabetes causes more morbidity and mortality than PBC [12,13]. In fact, type 2 diabetes is one of the leading causes of premature morbidity and mortality [20]. It has been shown that the main impact on QOL in PBC is fatigue [4,5]. Itching is also an important symptom in PBC that negatively affects patient QOL [21]. As the rate of progression of PBC has been slowed by ursodeoxycholic acid (UDCA) therapy [22], patient concerns about the risk of disease progression to cirrhosis and its complications have been much reduced so that most of their concerns are now about the symptom burden with its adverse impact on quality of life [23]. In comparison with autoimmune hepatitis, fatigue seems more pronounced and specific in PBC [24]. Fatigue is also an important clinical complaint of patients with type 2 diabetes, particularly women [25,26]. Further studies might investigate which physical domains of QOL are impacted differently by PBC and diabetes, as the scale used in this study was very short and general and did not provide such information.

Regarding illness perception, the results of this study for diabetes are close to those found for women and men with the same disease in New Zealand [17]. As hypothesized, there were several differences between PBC and diabetes. First, women with PBC reported feeling more symptoms related to their disease compared to women with diabetes. This is consistent with the result for physical QOL. Differences were also observed for *timeline* and *concern*. In

Table 2 Quality of life and illness perception by disease.

	Total	PBC Entire sample		PBC Without cirrhosis		Diabetes		PBC entire sample vs. diabetes		PCB without cirrhosis vs. diabetes	
	Mean (SD)	Mean (SD)	95% CI	Mean (SD)	95% CI	Mean (SD)	95% CI	F	P value	F	P value
Quality of life (SF-12)											
Physical QOL	41.4 (9.9)	39.8 (8.6)	38.4—41.3	39.9 (8.7)	38.2—41.5	44.7 (11.6)	41.9—47.6	11.458	0.001	10.199	0.002
Mental QOL	40.2 (5.8)	40.1 (6.0)	39.0—41.1	40.2 (6.1)	39.0—41.3	40.3 (5.3)	39.0—41.5	0.687	0.408	0.847	0.359
Illness perception (Brief-IPQ)											
Consequences	4.6 (2.7)	5.1 (2.7)	4.7—5.6	5.1 (2.7)	4.6—5.5	4.0 (2.6)	3.3—4.5	3.483	0.064	2.996	0.085
Timeline	9.6 (1.5)	9.9 (0.7)	9.7—10	9.9 (0.7)	9.7—10	9.1 (2.4)	8.4—9.7	7.780	0.006	7.224	0.008
Personal control	5.6 (2.8)	5.3 (2.9)	4.9—5.8	5.4 (2.9)	4.9—5.9	6.1 (2.3)	5.6—6.8	3.580	0.060	2.832	0.094
Treatment control	8.3 (2.3)	8.2 (2.4)	7.7—8.5	8.2 (2.4)	7.7—8.6	8.5 (2.0)	8.0—9.0	1.428	0.234	1.229	0.269
Identity	3.8 (2.8)	4.3 (2.8)	3.8—4.8	4.3 (2.8)	3.7—4.7	2.8 (2.6)	2.1—3.4	7.962	0.005	7.968	0.005
Concern	7.5 (2.8)	8.0 (2.4)	7.6—8.4	8.1 (2.4)	7.6—8.5	6.5 (2.2)	5.6—7.4	5.340	0.022	6.066	0.015
Understanding	7.2 (2.5)	7.1 (2.7)	6.6—7.5	7.0 (2.7)	6.5—7.5	7.4 (2.2)	6.9—8.0	0.441	0.507	0.499	0.481
Emotional response	5.5 (3.1)	6.0 (2.8)	5.0—5.9	6.0 (2.8)	5.5—6.6	4.5 (3.5)	3.6—5.3	3.153	0.077	3.603	0.059

P values are given after adjustment for age and disease duration.

Table 3 Illness perception item ''*causes*'' by disease.

	Total	PBC Entire sample	PBC without cirrhosis	Diabetes
Causes (%)				
Genetics	31.7	29.7	29.7	36.1
Emotion	22.8	30.5	30.5	6.6
Lifestyle	13.2	0.8	0.8	39.3
Unknown or unlucky	12.7	18.0	19.5	1.6
Medical	12.2	15.6	15.3	4.9
Age	7.4	5.5	4.2	11.5

PBC entire sample vs. Diabetes: Chi^2 = 71.936 and $P < 0.001$. PBC without cirrhosis vs. Diabetes: Chi^2 = 69.874 and $P < 0.001$.

both diseases, women felt that their illness would continue almost forever, as *timeline* scores were very high in both groups. This is not surprising as both diseases are chronic. However, this feeling was significantly greater in PBC. Moreover, PBC women felt more concerned by their illness than women with diabetes. These results might be explained by the fact that PBC is a rare disease, which could lead to a more pessimistic perception and a burning sense of injustice [14]. This hypothesis could also explain the marginally significant differences observed: women with PBC reported that illness affected their life more (*consequences*), that they had a lower personal control over their illness (*personal control*) and felt more affected emotionally (*emotional response*) than women with diabetes. Patients with type 2 diabetes can control their illness by using health behaviors. This has been shown specifically in studies focusing on the self-management of glycemic control [27], which might give a stronger feeling of control over the illness. The results dealing with emotional responses can be related to research on emotions in patients with chronic diseases. Anxiety and depression have been particularly studied. In diabetes, anxiety and depression symptoms seem much higher than in the general population, around 32% and 24%, respectively [28], while clinical depression is around 14.1% [29]. In PBC, earlier studies observed a high prevalence of depression, at around 50% [30]. A more recent study found that major depression is around 4.2%, which is lower than in the general population, although many patients report fatigue and other somatic symptoms such as loss of weight, insomnia and loss of concentration [31]. The prevalence of anxiety has not been specifically studied in PBC. These results suggest that diabetes patients report more clinical depression than PBC patients whereas the results of our study underline that, in relation to their disease, women with PBC feel more concerned and affected emotionally. It would be interesting to explore more precisely these emotions in relation to disease. It has been shown that negative emotions, mainly frustration, guilt, anger and anxiety, are especially reported by women having PBC with fatigue [32]. However, as our results were not significant ($0.05 < P < 0.08$), they should be taken with cautious and further research is needed on emotional aspects.

The results also showed that, in both diseases, women reported that their treatment greatly helped their illness (*treatment control*) and that they clearly understood their illness (*understanding*). Regarding *treatment control*, this may be explained by the fact that the two diseases have established therapies, which are effective if the patient is adherent. The results also underline that having a rare disease such as PBC does not induce less understanding than having a common disease such as diabetes.

Finally, cirrhosis is known to affect quality of life [33]. In our PBC sample, few women reported a stage IV disease (5.1%) and significant differences between women with PBC and women with diabetes did not change when women with cirrhosis were excluded. These results are in line with those from Poupon et al. [3] who found no associations between quality of life and histological stages in PBC.

Two main limitations of our study warrant mention. The first is that the samples were not selected randomly and therefore the results may be subject to bias. In addition, the recruitment methods were not the same (through the Hepatology Unit and patient association for the PBC sample and through general practitioners for the diabetes sample), which might have influenced the results. People who belong to a patient association probably question themselves more about their disease, and try to understand it better in order to deal with their worries. A second limitation is the PBC diagnosis, which was a self-reported diagnosis for women recruited through the patient association. Consequently, the results should be viewed with caution and further studies are needed to confirm the differences observed.

Regarding the clinical implications, the results of this study underline the need for health professionals to be sensitive to patient QOL and illness perception in PBC. Physician empathy, particularly concerning patient illness representations, can have an impact on various patient outcomes [34]. Moreover, as supported by Leventhal's Self-regulation Model [35], illness perception in diabetes may have direct and independent effects on both dietary self-care and diabetes distress [36]. Interventions focusing on illness perception could be developed to try to enhance the patient's sense of control, helping individuals to deal with emotions related to the disease and feel the symptom burden less. This might enhance QOL.

To conclude, this study provides insights into the quality of life and illness perception of women with PBC. Further studies are needed to understand PBC specificities better, especially regarding physical QOL. Research might also investigate how illness perception evolves during PBC and how it affects QOL in longitudinal studies.

Disclosure of interest

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