

The Illness and Treatment Beliefs of Adults with Blepharospasm and Hemifacial Spasm Receiving Botulinum Toxin Injections: A Cross-Sectional Study

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Abstract

Background/ Aims: Benign essential blepharospasm (BEB) and hemifacial spasm (HFS) cause eyelid and/or facial spasms. Treatment with botulinum toxin leads to a cyclical pattern of relief followed by symptoms returning. Limited evidence suggests a link between patients' beliefs about their symptoms and perceived efficacy of treatment but further research is needed to establish the specific beliefs held by BEB and HFS about their condition and its treatment.

Methods: A cross-sectional study of 130 patients with BEB or HFS was conducted at Moorfields Eye Hospital, London. Beliefs about the conditions and treatments were assessed using the Illness Perceptions Questionnaire (IPQ-R) and Treatment Representations Inventory (TRI). Patients also completed the Blepharospasm Disability Index (BSDI) and Craniocervical Dystonia Questionnaire (CDQ-24). Severity and frequency of spasms were recorded using the Jankovic Rating Scale (JRS). Relationships between the outcomes were identified using Pearson's product-moment correlation.

Results: A range of beliefs about the conditions and botulinum toxin were identified: participants associated BEB and HFS with a chronic timeline and believed their condition was not caused by psychological factors, risks, or their personality or lifestyle. Participants also demonstrated decision satisfaction related to treatment but had some treatment concerns. There was notable uncertainty in a number of patients' beliefs.

Conclusions: Participants believed their condition to be chronic and generally understood what caused BEB or HFS. Strong relationships between patients' beliefs about their condition and treatment, and aspects of quality of life, were identified. This study highlights the uncertainty patients face in managing their condition which could be addressed in future interventions to improve patient outcomes, including quality of life.

Background

Benign essential blepharospasm (BEB) and hemifacial spasm (HFS) are movement disorders characterised by involuntary closure of both eyelids (BEB) or spasms affecting one side of the face (HFS). Adults with BEB and HFS report poorer health-related quality of life than unaffected controls [1]. Although no differences have been found between the two populations for generic quality of life outcomes [2], patients with BEB have been found to have poorer vision-related quality of life [3].

Botulinum toxin (BTX) is considered the most effective treatment for BEB and HFS [4] and is used to alleviate spasms. However its effects do not last, resulting in a fluctuating pattern of relief and symptom return [5]. A treatment regimen of standard treatment intervals, usually every 3 months, is commonly used despite patients reporting varying periods of treatment effectiveness [5]. This treatment experience, whereby some patients may be left experiencing debilitating symptoms until their next scheduled appointment while others are seen too often, could impact on beliefs about treatment. Initial evidence suggests beliefs about treatment effectiveness can drive BTX discontinuation. A ten-year follow-up study

of BTX suggested a “honeymoon” period whereby initial injections were perceived to be more effective than subsequent injections, resulting in some patients deciding to discontinue BTX regimen altogether [6]. Understanding patients’ beliefs about their condition and treatment is pertinent to our ability to explain treatment adherence, healthcare use, and quality of life.

A growing body of research has explored patients’ beliefs about a number of long-term conditions [7, 8] and their treatments [9]. Including how these are linked to psychosocial and behavioural outcomes, such as coping [10, 11], mood [8, 12, 13], quality of life [8] and treatment adherence [14, 15]. Through examining these beliefs, a greater understanding of what drives adjustment and behaviours, including adherence to medications and healthcare resource use, can be established. There has however been scarce coverage of this topic in BEB and HFS research.

Research in other movement disorders, including Parkinson’s Disease (PD), has identified links between poorer psychological adjustment and perceptions about symptoms related to the condition, that conditions have a cyclical nature, that conditions lead to serious and negative consequences, feeling a lack of control and understanding over the condition, and negative emotions about the condition [11, 16]. These findings indicate the important role of beliefs in understanding the process of adjusting to living with a movement disorder. Further understanding in BEB and HFS may help explain differences in treatment adherence, quality of life, and be used to design future interventions to improve wellbeing and illness self-management. This study aimed to identify patients’ beliefs about BEB and HFS and their treatment and to examine how these beliefs relate to clinical and psychological outcomes, so future interventions can be tailored to resolve patients’ concerns and improve outcomes.

Materials And Methods

Aim

To identify the beliefs patients with BEB and HFS hold about their condition and treatment and to identify the relationships between these beliefs and clinical and psychosocial outcomes.

Study design

This cross-sectional study used baseline data from a single-masked randomised controlled trial (RCT) assessing the effectiveness of a patient-initiated BTX service compared to standard care [17].

Setting

This study was conducted in the nurse-led BTX clinic at Moorfields Eye Hospital (MEH) NHS Foundation Trust, London.

Participants

Participants were recruited from the nurse-led BTX clinic at MEH London. The hospital records of patients booked for the BTX clinic were screened by a research nurse for eligibility. Eligibility criteria included adults (>18 years of age) with a diagnosis of BEB or HFS, two stable doses of BTX previously, capacity to give informed consent, and able to communicate fluently in written and/or spoken English. Although BEB and HFS are different conditions they share many characteristics, including procedural aspects of the treatment and the cyclical nature of the response, hence including both in this research.

Ethical approval

Ethical approval for this study was obtained from the National Research Ethics Service, London – Queen Square (REC reference 15/LO/0439) in April 2015.

Procedure

Participants meeting the inclusion criteria were invited by letter to participate in the RCT two weeks before their clinic appointment, with an information sheet attached. Patients were approached by a member of the clinical team at their appointment to confirm if they wished to take part. Consenting participants received a questionnaire booklet with a freepost return envelope and completed the questionnaire at their clinic visit or at home.

Materials

The questionnaire pack asked for demographic information: age, gender, marital status, housing situation, ethnicity, employment status, and education level. Clinical measures were recorded by the treating clinician including diagnosis, duration of BTX treatment, number of previous cycles, and frequency of previous cycles.

Severity and frequency of spasms

BEB or HFS severity was measured using the Jankovic Rating Scale (JRS) [18], consisting of two subscales measuring severity and frequency of spasms and found to be valid and reliable in BEB [19]. For patients with HFS an additional rating scale measuring the severity and frequency of cheek involvement developed previously for HFS research [20] was used. Higher scores indicate severe and more frequent spasms, with a maximum total score of 8 for each rating scale

Illness disability

The Blepharospasm Disability Index (BSDI)© [21] was used to measure disability when performing six daily activities: reading, driving, watching TV, shopping, walking and doing everyday activities. Responses ranged from “no impairment” to “no longer possible due to illness”, with a “not applicable” option available for each item and higher scores indicating greater disability. This valid and reliable measure [19] is also recommended for use with patients with HFS [20].

Illness beliefs

Illness perceptions were measured using the Revised Illness Perceptions Questionnaire (IPQ-R), which is divided into 3 sections. The identity subscale is presented as a list of 14 common symptoms [7], e.g. fatigue. Following recommendations by the original authors, four symptoms related to BEB and HFS were added: (i) frequent blinking, (ii) irritation of the eye, (iii) uncontrollable eye closure and, (iv) muscle twitching. Respondents indicate whether they have experienced each symptom and whether symptoms are related to their condition. The maximum score was 18, with higher scores demonstrating a belief that more symptoms are associated with the condition.

The original IPQ-R contained 38 items in the second section. The factor structure, validated in a sample of patients with eight illnesses, included *timeline acute/ chronic*, *timeline cyclical*, *consequences*, *personal control*, *treatment control*, *illness coherence* and *emotional representations* [7]. Patients with BEB and HFS were not included in the original validation, so to capture the specific beliefs of these patients, construct validity was assessed by the authors using a principal component analysis. The 38 IPQ-R items were found to be structured by 10 components in this sample of patients with BEB and HFS: *timeline acute*, *timeline chronic*, *timeline cyclical*, *consequences*, *positive personal control*, *negative personal control*, *illness uncertainty*, *treatment control*, *illness coherence* and *emotional representations*. Higher scores represent strongly held beliefs about chronicity, the cyclical nature of BEB or HFS, their negative consequences, and negative emotional responses towards the conditions. Higher scores on positive personal control, treatment control or coherence dimensions represent positive beliefs about controllability of BEB or HFS and their treatment and a personal understanding of their illness. Higher scores on the negative personal control and uncertainty domains indicate beliefs in being unable to control the conditions and that they cause uncertainty.

The third section measuring causes includes 18 items. A factor analysis revealed four types of causal beliefs held by the eight illness groups assessed for the original questionnaire development: *psychological attributions*, *risk factors*, *immunity*, and *accident or chance* [7]. These items were also tested by the authors for construct validity in the current sample of patients with BEB and HFS. Four constructs were identified: *psychological attributions*, *risk factors*, *lifestyle*, and *chance*. Higher scores on each subscale indicates stronger beliefs the condition was caused by these factors.

The internal consistency of the new IPQ-R subscales was found to be high (Table 2), with the exception of the chance subscale ($\alpha=0.18$). When the item relating to “accident or injury” was removed a satisfactory Cronbach’s alpha of 0.50 was achieved for this subscale (Table 2).

Treatment beliefs

Beliefs about BTX were measured using the Treatment Representations Inventory (TRI) [9]. This 27-item measure consists of four scales measuring treatment value, treatment concerns, decision satisfaction and cure, with higher scores representing stronger beliefs in these areas. This measure demonstrated good internal consistency in the current sample when tested by the authors, with Cronbach's alphas ranging from 0.67 for the cure subscale to 0.86 for the decision satisfaction subscale (Table 2).

Quality of life

Quality of life was measured using the Craniocervical dystonia questionnaire (CDQ-24;[22]), which has been used successfully in BEB and HFS [23]. The CDQ-24 assesses adjustment in five life domains: emotional well-being, activities of daily living, social/family life, pain, and stigma. Raw subscale scores were linearly transformed to a 0-100 scale, with higher scores indicating poorer quality of life.

Statistical analysis

All analyses were undertaken using IBM SPSS Statistics Version 22.0.

Missing data

Missing data was dealt with using mean imputation methods. Where over 50% item-level data were missing the subscale score was considered missing. If over 50% total data were missing for a single participant their data were not analysed.

Relationships between factors

Correlations between illness perceptions (IPQ-R), treatment perceptions (TRI), quality of life (CDQ-24), disease severity (JRS), and disability (BSDI) were assessed using Pearson's product-moment correlation. Where data were not normally distributed, Spearman's rho was used.

Group differences

Comparisons between the beliefs of patients with BEB and HFS were made using independent samples *t* tests. Non-parametric Mann Whitney U tests were used for any subscale that was found to be non-normally distributed.

Results

A total of 247 patients screened by the nurse were eligible and invited to take part in the study. Five eligible patients did not attend a clinic appointment during the recruitment period and 87 declined to participate when approached. A total of 155 patients provided written consent, 130 of whom returned a baseline questionnaire and were included in the study.

Missing data

One participant did not complete over 50% of their baseline questionnaire and their data were removed from the analysis. All scales demonstrated less than 10% data missing and mean imputation was used to minimise the impact of missing data [24].

Participant characteristics

The characteristics of patients included in the study can be found in Table 1.

Table 1. Participants' demographic characteristics, severity, and disability of BEB and HFS

Variable	Number	Median	Interquartile Range	Range	Mean \pm SD
Age (years)	37 (29%)	1.0	0.33-1.83	37-88	63.84 \pm 10.8
Gender				4-336	
<i>Male</i>	92 (71%)			2-121	81.4 \pm 71.8
<i>Female</i>	89 (69%)			1-9	21.5 \pm 21.1
Ethnicity					
<i>White British/Irish/Other</i>	40 (31%)				3.19 \pm 1.2
<i>Black African/Caribbean/Asian/Other</i>	76 (59%)				
Diagnosis	53 (41%)				
<i>BEB</i>					
<i>HFS</i>					
Duration of BTX (months)					
Number of pervious cycles					
Usual time between treatments (months)					
Disability (BSDI)					
Severity (JRS)					
<i>Eyelid Spasms</i>		2.0	0-5		
<i>Cheek Involvement (HFS only)</i>		4.0	2-5.5		

Illness beliefs

Mean illness identity scores demonstrated patients experienced few symptoms attributed to BEB or HFS (Table 2). Of symptoms common to BEB and HFS, just over half of participants felt frequent blinking (56%) was related to their BEB or HFS. Similar proportions of the sample attributed sore eyes (58%), eye irritation (68%), uncontrollable eye closure (67%) and muscle twitching (67%) to their BEB or HFS. Between 30 and 45% of the sample also attributed sleep difficulties, headaches, fatigue and pain to their condition.

The mean scores for IPQ-R subscales (Table 2) demonstrate participants held strong beliefs that BEB and HFS are chronic rather than acute in their timeline and that treatment generally controlled their condition. Participants generally did not believe their BEB and HFS to be caused by psychological, risk, or personal lifestyle factors. Participants demonstrated uncertainty in a number of their beliefs (Figure 1).

[INSERT FIGURE 1 HERE]

Treatment beliefs

TRI subscale scores revealed particularly strong satisfaction among patients with BEB and HFS with the decision to undergo BTX therapy, along with some general uncertainty about their treatment (Figure 1).

Correlations

When BEB or HFS was believed to be associated with more negative consequences (IPQ-R consequences), significant associations were found with difficulties in activities

Table 2. Participants' scores on measures of illness and treatment beliefs and differences in scores between BEB and HFS

IPQ-R & TRI Subscale	Items	Internal consistency (Cronbach's α)	Total sample Mean (SD)	Mean Score (SE)	BEB Mean (SD)	HFS Mean (SD)	Statistical differences between BEB and HFS
IPQ-R Identity	18	.80	5.28 (3.27)	5.31 (0.30)	6.48 (3.09)	4.41 (2.84)*	p = 0.001
IPQ-R Acute timeline	3	.68	10.80 (2.60)	3.59 (0.08)	10.94 (2.44)	10.74 (2.86)	p = 0.596
IPQ-R Chronic timeline	4	.88	16.74 (3.57)	4.15 (0.08)	16.90 (3.42)	16.50 (3.80)	p = 0.541
IPQ-R Consequences	6	.85	19.34 (5.33)	3.23 (0.08)	20.61 (5.20)	17.30 (4.88)**	p < 0.001
IPQ-R Negative personal control	2	.70	5.53 (2.13)	2.78 (0.09)	5.72 (2.18)	5.29 (2.07)	p = 0.227
IPQ-R Positive personal control	4	.77	9.46 (3.44)	2.37 (0.08)	9.52 (3.48)	9.38 (3.44)	p = 0.777
IPQ-R Treatment control	3	.67	11.37 (2.12)	3.79 (0.06)	11.30 (1.98)	11.56 (2.25)	p = 0.558
IPQ-R Coherence	5	.86	14.09 (4.58)	2.81 (0.08)	13.59 (4.19)	14.77 (4.95)	p = 0.160
IPQ-R Cyclical timeline	2	.78	6.33 (1.98)	3.17 (0.09)	6.25 (2.01)	6.46 (1.94)	p = 0.454
IPQ-R Emotional reps	5	.89	14.41 (5.04)	2.88 (0.09)	15.00 (4.93)	13.62 (5.12)	p = 0.136
IPQR Illness Uncertainty	3	.60	9.34 (2.49)	3.12 (0.08)	9.54 (2.36)	9.02 (2.64)	p = 0.259
IPQ-R Cause – psychological attributions	5	.86	12.25 (4.92)	2.44 (0.09)	13.30 (4.87)	10.86 (4.71)*	p = 0.004
IPQ-R Cause – risk factors	6	.77	12.30 (4.10)	2.04 (0.06)	12.96 (3.94)	11.32 (4.28)*	p = 0.035
IPQ-R Cause - lifestyle	4	.70	6.91 (2.56)	1.72 (0.06)	6.74 (2.47)	7.00 (2.69)	p = 0.806
IPQ-R Cause-chance	2	.50	5.59 (2.04)	2.77 (0.09)	5.68 (1.98)	5.44 (2.07)	p = 0.320
TRI Treatment value	5	.79	16.00 (4.45)	3.19 (0.08)	16.46 (4.36)	15.20 (4.64)	p = 0.141

TRI Treatment concerns	6	.84	15.99 (5.12)	2.66 (0.08)	16.06 (5.05)	15.53 (4.98)	p = 0.575
TRI Decision satisfaction	9	.86	35.42 (5.23)	3.93 (0.05)	34.96(4.49)	36.06 (5.82)	p = 0.416
TRI Cure	7	.67	21.99 (4.39)	3.14 (0.06)	21.42 (3.82)	22.85 (4.95)	p = 0.080

of daily living (CDQ24 $r = 0.62$ $p < 0.001$), poorer overall quality of life (CDQ-24 $r = 0.67$ $p < 0.001$), and greater disease-related disability ($r = 0.53$; $p < 0.001$). A higher illness identity (IPQ-R Identity) was significantly associated with more difficulties in activities of daily living (CDQ-24) $r = 0.50$ ($p < 0.001$) and greater levels of disability $r = 0.37$ ($p < 0.001$). Poorer emotional wellbeing (CDQ24) was strongly associated with holding more negative emotions about the condition (IPQ-R emotional representations) $r = 0.70$ ($p < 0.001$). Higher levels of disease disability were also associated with strongly held beliefs that treatment would not cure the condition $r = -0.20$ ($p < 0.05$) and placing greater value on the BTX treatment $r = 0.23$ ($p < 0.05$), although these associations were weaker. Of note, few significant relationships were found between severity and frequency of spasms, and illness and treatment beliefs. More frequent and severe eyelid spasms were associated with more value being placed on treatment $r = 0.20$ ($p < 0.05$) and more negative consequences as a result of the condition $r = 0.19$ ($p < 0.05$), although these were both weak relationships. The full results of relationships between the variables is reported in Supplementary File 1.

There were a number of significant relationships between the perceptions patients held about their illness, and their perceptions of their treatment. A belief that treatment can cure the condition was moderately associated with believing the condition is less chronic in nature $r = -0.37$ ($p < 0.001$). More value being placed on treatment was strongly associated with more negative emotional representations $r = 0.48$ ($p < 0.001$), and experiencing more negative consequences associated with the condition $r = 0.5$ ($p < 0.001$).

Differences between BEB & HFS

A number of differences in beliefs were found between BEB and HFS participants (Table 2). Patients with BEB associated more symptoms with their condition, viewed their condition as having greater negative consequences and were also more likely to believe their condition was caused by psychological factors, such as stress, and risk factors, such as altered immunity, than patients with HFS.

Discussion

This study aimed to identify the illness and treatment beliefs held by adults with BEB and HFS and how these were related to clinical factors and psychological adjustment. This study found strong beliefs among participants about the chronicity of their condition, supporting findings in PD [16]. This study demonstrates patients with movement disorders affecting the face and/or eyes also understand their condition is serious and long-term. Beliefs about more symptoms associated with BEB, and more

negative consequences of BEB, than HFS, may be due to the greater likelihood for visual impairment when spasms affect both eyes (BEB) compared to one eye (HFS), a suggestion supported by previous research [20].

Participants associated common BEB and HFS symptoms with their condition. However patients in this study identified additional symptoms not normally associated with dystonia, including fatigue, pain and sleeping difficulties. These same symptoms strongly predict depression in patients with chronic conditions, including rheumatoid arthritis [25]. This suggests further research exploring depression in BEB and HFS would be useful to inform the clinical management of these chronic conditions.

With evidence suggesting that the common cause of BEB and HFS is neurological [19, 20], participants generally demonstrated a coherent understanding of what caused their condition. This is an important issue because causal beliefs can influence the treatment patients seek and, ultimately, how successfully they manage their condition [26]. Patients with BEB were more likely to associate their condition with psychological or risk factors than those with HFS. There was also uncertainty among participants about whether their condition was down to chance. Patients could be better informed about the causes of BEB and HFS to resolve any uncertainty or misunderstanding.

Participants with both BEB and HFS were satisfied with their decision to undergo BTX, supporting previous findings [27]. Satisfaction with treatment decisions can impact treatment-related behaviours, including medication adherence [28]. In the context of BEB and HFS, satisfaction may have an impact on whether patients decide to continue treatment. This study also found associating BEB and HFS with more negative consequences and emotions was linked to placing more value on BTX. This is further emphasised by a general belief among patients in this study that their treatment controlled their condition, demonstrating the importance of treatment to patients with BEB and HFS.

Strong relationships between patients' beliefs about their condition and treatment, and quality of life, were identified. A recent meta-analysis also found perceptions of symptoms, illness consequences and emotional responses were consistently related to psychosocial outcomes for a range of chronic conditions, including PD [8]. This suggesting patients' perceptions of BEB and HFS, and the impact of these perceptions, are comparable to other chronic conditions which are demanding and involve making life adjustments. These relationships were stronger than relationships between beliefs and clinical factors, suggesting the clinical severity or physical disability associated with BEB and HFS are not helpful predictors of patients' personal appraisals of their condition and treatment.

Noteworthy is the uncertainty patients felt in several of their beliefs. Participants did not appear certain their condition is cyclical, that it is associated with negative emotions, or that they understood their condition. Participants did not appear certain about the value of their BTX treatment or whether this was curative. Uncertainty is important in long term conditions without a known cause or cure [29] as it has been linked to poor quality of life across a range of chronic conditions [30]. BEB and HFS involve fluctuating patterns of severity and unpredictable symptoms. Coupled with feeling uncertain about

whether or not treatment is curative, patients could struggle with choosing appropriate coping strategies or managing the condition effectively [31].

As this study was cross-sectional it is not possible to draw inferences about the direction of causality. To understand the predictability of illness and treatment beliefs on patient-related outcomes, such as quality of life, longitudinal data are required. The applicability of these findings outside of the UK health system is also limiting. Treatments would likely be of different value to patients depending on whether they are administered frequently or infrequently, and whether they are free or costly.

Conclusions

Adults with BEB and HFS held strong beliefs about the chronic timeline of their condition and that BTX treatment was the most suitable option for them. Participants generally had a coherent understanding of the cause of BEB and HFS. However patients demonstrated uncertainty in several beliefs about their condition and treatment. Associations between negative illness and treatment beliefs, and difficulties in daily living, poor emotional wellbeing and poorer overall quality of life have been identified in this study, which require further exploration in longitudinal research. Future interventions could directly target patients' beliefs about their BEB and HFS and their treatment, and address any uncertainties, to improve successful management of these debilitating conditions and patient outcomes.

Abbreviations

BEB	Benign Essential Blepharospasm
BSDI	Blepharospasm Disability Index
BTX	Botulinum toxin
CDQ	Craniocervical Dystonia Questionnaire
HFS	Hemifacial Spasm
IPQR	Illness Perceptions Questionnaire - Revised
JRS	Jankovic Rating Scale
MEH	Moorfields Eye Hospital NHS Foundation Trust
NHS	National Health Service
PD	Parkinson's Disease
RCT	Randomized controlled trial
REC	Research Ethics Committee
SD	Standard deviation
SPSS	Statistical Package for Social Sciences
TRI	Treatment Representations Inventory
UK	United Kingdom

Declarations

Ethical approval

Ethical approval for this study was obtained from the National Research Ethics Service, London – Queen Square (REC reference 15/LO/0439) in April 2015.

Ethics approval and consent to participate

Full NHS Research Ethics Committee (REC) approval was received (REC reference: 15/LO/0439) from London – Queen Square REC. All participants provided written consent to participate.

Consent for publication

The manuscript does not contain any individual's data requiring consent.

Availability of data and materials

Anonymized data from the current study can be made available, if approved by our REC.

Competing interests

No conflict of interest exists for any author.

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Author's contributions

DGE (principal investigator), SPN and HM conceived the study. DGE, SPN, HM, SLW, SPN, CH, ND, DS and BC designed the study. SLW, ND and DGE recruited participants and collected the study data. SLW and SPH analyzed the data. SLW drafted the manuscript. All authors revised the manuscript for intellectual content and approved the final manuscript.

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Figures

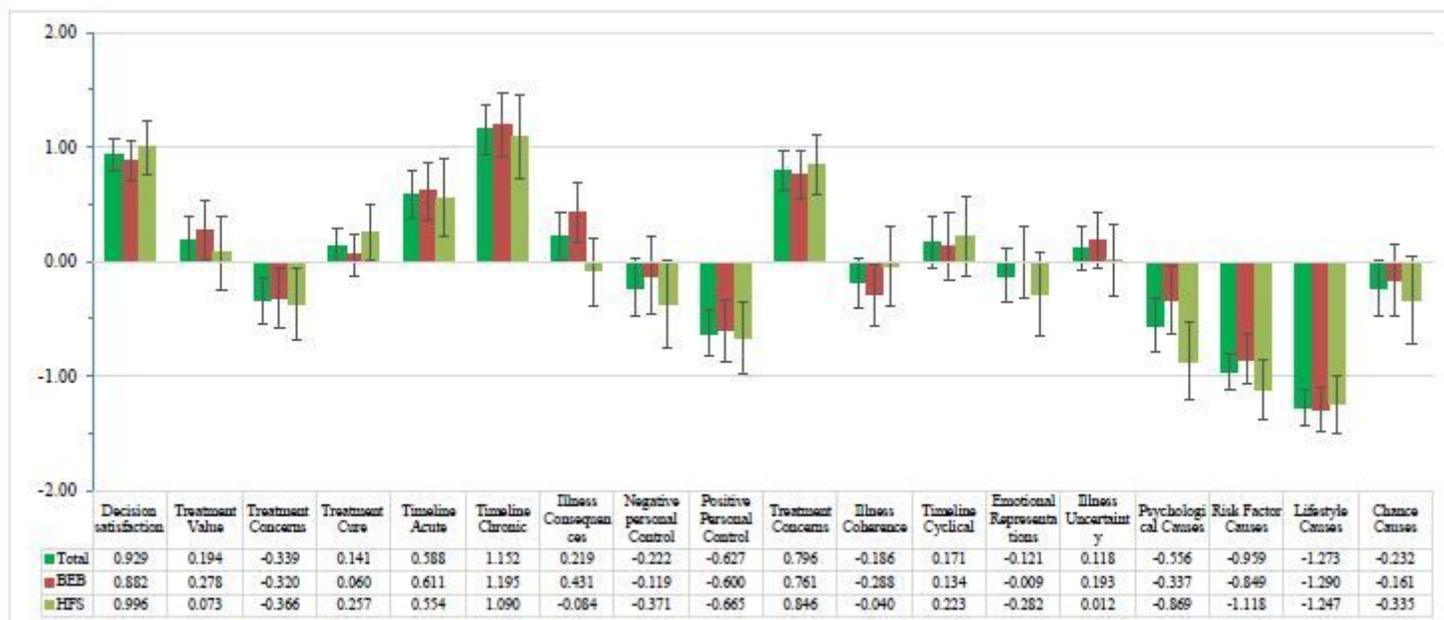


Figure 1

Mean illness and treatment belief scores • y Axis: Mean IPQR and TRI scores • x Axis: IPQR and TRI subscales