Haemophilic Arthropathy and Health-Related Quality of Life in Haemophilic Patients

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Introduction

In severe haemophilia A patients, more than 80% of the spontaneous bleeding events occurring in the joints and muscles. Repeated joint bleeding leads to chronic synovitis, cartilage damage and bony destruction, which results in reduced range of motion (ROM), painful joints and muscle atrophy. Haemophilic arthropathy can negatively influence perception of health-related quality of life (HRQOL) in haemophiliacs.

HRQOL is an important outcome in the care of the patients of haemophilia. Many clinical variables have been found associated with lower HRQOL in hemophilia including disease severity, bleeding, chronic pain, presence of blood-borne virus infection (Human Immunodeficiency Virus and hepatitis C virus), and orthopaedic status. To the best of our knowledge, reports on the HRQOL of the hemophilia patient in Taiwan are not available.

The aims of our study were (1) to evaluate HRQOL of hemophilia patients in Taiwan (2) to determine the incidence of hemophilic arthropathy in our cohort of 86 hemophiliacs and (3) to comparatively correlate HRQOL with clinical factors of hemophilia patients.

Materials and methods

A total of 86 patients who had been followed at our haemophilia center were studied. We collected clinical information including age, haemophilia type, disease severity, factor inhibitor, HBV, HCV, HIV, liver function and SF-36. Bilateral shoulders, elbows, hips, knees and ankles were evaluated in terms of range of motion and Pettersson score by X-ray on the same day. The relationships between SF-36 and clinical variables were assessed using Spearman's correlation coefficient.

Results

Eighty-one hemophilia A and 5 haemophilia B patients were enrolled. The mean age was 30.64±14.1 years (range, 6 to 66 years). There were four HBV (4.7%), 5 HIV (5.8%) and 46 HCV (53.5%) carriers in the 86 patients. (Table 1) In haemophilic arthropathy, the most common affected joint was ankle (57 patients, 66.3%) followed by elbow (46 patients, 53.5%) and knee (41 patients, 47.7%).(Fig. 1) The SF-36 scores of haemophilic patients were worse in comparison with Taiwanese normative values. (Fig. 2,3) The significant correlations between the summary score of SF-36 and Pettersson score(r=-0.560, p<0.001), ROM(r=0.538, p<0.001), age (r=-0.426, p<0.001) and HCV (r=-0.356, p=0.001) were noted. (Table 2) There was no significant correlation between other clinical variables and SF-36.

Demographic features

Characteristic	Average	Range
Patient number	86	(A:81 B:5)
Age, y/o,	30.64±14.1	6-66
Height (cm)	164.95±13.4	120-183
Body weight (kg)	65.33±17.8	20.4-108.3
BMI (kg/m²)	23.62±5.1	13.2-38.8
Severity		
severe	59	68.6%
moderate	15	17.4%
mild	12	14.0%
Inhibitor	6	
HIV	5	
HBV	4	
HCV	46	
Short Form 36	65.61±20.17	16-98
Pettersson score of joints*	28.12±28.6	0-102

Table 1. Demographic features

Correlations between QOL and clinical variables

Pettersson score	r= -0.560	P<0.001
ROM	r= 0.538	P<0.001
Age	r= -0.426	P<0.001
HCV	r= -0.356	p=0.001
Severity	r= -0.113	P=0.299
HIV	r= -0.076	P=0.485
HBV	r= -0.008	P=0.943
GPT	r= -0.013	P=0.663
Inhibitor	r= -0.092	P=0.399

Table 2. Correlations between HRQOL and clinical variables

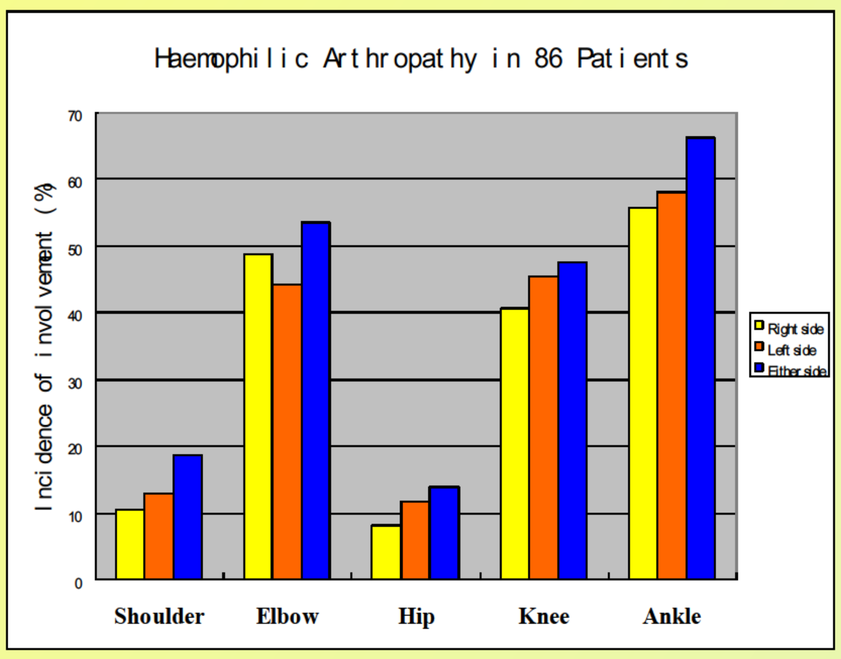

Fig 1 Haemophilic arthropathy

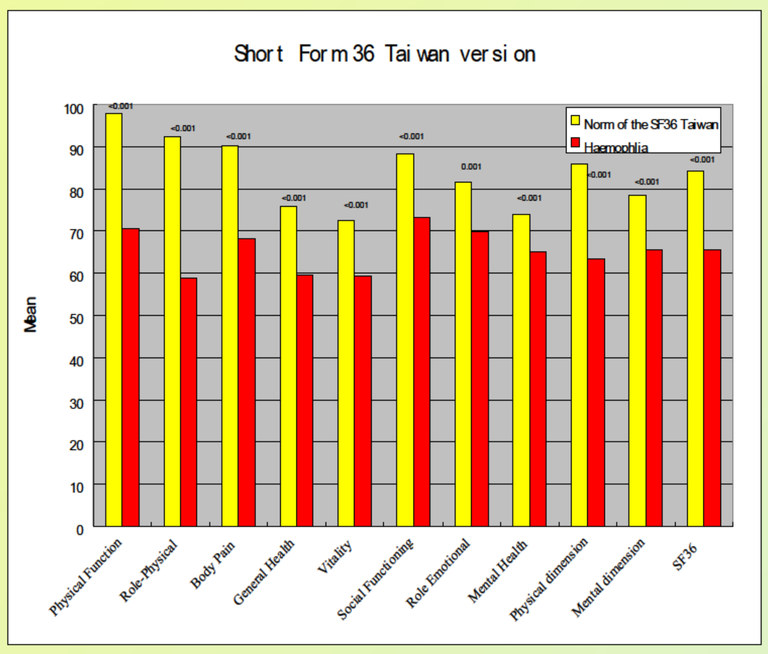

Fig 2. Short Form-36

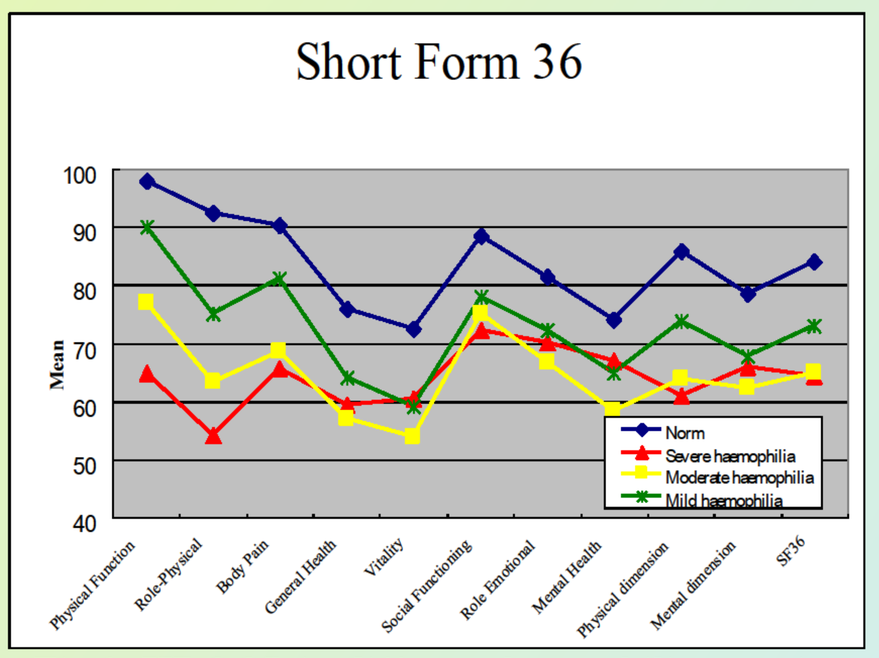

Fig 3. HRQOL in different hemophilia severity

Conclusion

Advanced Pettersson score, reduced ROM, older age and HCV infection were related to lower HRQOL in haemophilic patients. The worse the arthropathy is, the lower the HRQoL. If haemophilic arthropathy can be managed appropriately to prevent ROM deterioration, there is a substantial quality of life improvement for haemophiliacs.

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