



Outcomes of total knee arthroplasty in the adult Kashin-Beck disease with severe osteoarthritis

Zhan-Kui Jin¹ · Ying Yang² · Cui-Xiang Xu³ · Bo Yang¹ · Mikko J. Lammi⁴ · Yan-Hai Chang¹ · Ming Ling¹ · Xiong Guo⁴ · Xiao-Qing Wang^{1,2}

Received: 30 October 2017 / Accepted: 11 June 2018 / Published online: 3 July 2018
© SICOT aisbl 2018

Abstract

Purpose Kashin-Beck disease (KBD) is an endemic osteoarthropathy, and the severe knee pain and functional limitations were seriously affecting the quality of life in patients with end-stage KBD. We retrospectively evaluated the clinical outcomes and the quality of life in KBD patients with total knee arthroplasty (TKA).

Methods A total of 22 subjects (25 knees) suffered KBD with severe knee pain and underwent primary TKA. Knee pain was measured by visual analogue scale (VAS), and the knee function was evaluated by Knee Society Clinical Rating System Score (KSS). KBD Quality of Life (KBDQOL) was used to evaluate the quality of life in KBD patients before and after TKA.

Results There were no major complications after TKA. The levels of VAS score were obviously decreased in post-operation than that in pre-operation. The levels of KSS score were increased in one year after TKA compared with the pre-operative values, and it maintained a higher level on three years after TKA. The average KBDQOL score level of each domain in pre-operation and one and three years after TKA was increased accordingly. The average scores of physical function, activity limitation, support of society, mental health, and general health in one year after TKA were significantly higher than those in pre-operation.

Conclusions TKA can reduce knee pain, improve knee function, and improve the quality life in KBD patients. KBDQOL questionnaire may be a promising instrument for assessing the quality life in KBD patients.

Keywords Arthroplasty · Knee · Osteoarthritis · Kashin-Beck disease · Quality of life

Introduction

Kashin-Beck disease (KBD), commonly known as “big bone disease” in China, was first described in 1849 [1]. The investigating history of KBD was isolated by time, such as Russian

(1850–1950), Japanese (1905–1945), and Chinese (from 1950 until now) periods of investigation [1]. The changes of KBD disease in China can be divided into three stages. In the first stage, from 1949 to 1980s, this disease was prevalent in KBD endemic areas. In the second stage, from 1990 to 2000, the overall incidence of KBD was declining. In the third stage, from 2000 to 2007, KBD had subsided in most of KBD endemic areas [2]. At present, cases of children with KBD in China have been controlled in more than 80% in the endemic areas according to clinical assessment and acceptance criteria and in more than 90% according to X-ray diagnostic examination and testing standards [2]. Obviously, KBD in children has been under control in China.

However, many adult KBD patients, who left in endemic areas, were experiencing severe joint deformity, pain, movement disorder, and lameness, so they cannot take care of themselves. The patients can relieve pain by non-surgical methods, such as non-steroidal anti-inflammatory drugs (NSAIDs) [3], glucosamine, chondroitin sulfate, intra-articular injection of hyaluronic acid [4], herbal medicines,

✉ Xiao-Qing Wang
56481980@qq.com

¹ Department of Orthopaedics, Shaanxi Provincial People’s Hospital (the Affiliated Hospital of Xi’an Medical University), Xi’an 710068, China

² Xi’an Medical University, Xi’an 710021, China

³ Center of Shaanxi Provincial Clinical Laboratory, Shaanxi Provincial People’s Hospital (the Affiliated Hospital of Xi’an Medical University), Xi’an 710068, China

⁴ Institute of Endemic Diseases, School of Public Health of Health Science Center, Xi’an Jiaotong University, Key Laboratory of Trace Elements and Endemic Diseases, National Health and Family Planning Commission, Xi’an 710061, China

and physical therapy. Operations such as arthroscopy are an effective method. However, treatment of arthroscopy is difficult to improve in larger osteophytes and extensive cartilage injuries in the joint and cannot significantly increase knee motion post-operatively. Therefore, joint replacement is still the most effective method for the end-stage KBD patients [5, 6] who suffered severe varus or valgus deformity, flexion deformity, and rotation mixed deformity in knee joint. However, a rare study has been published to date on joint replacement therapy in patients with end-stage KBD. Just three studies had analyzed total knee arthroplasty (TKA) [5, 7] and total hip replacement [6] in KBD patients, respectively, and showed excellent clinical and radiographic outcomes in short-term follow-up.

The quality of life in the patients is one of the key factors in assessing the success of the operation. There are several methods that have been used to assess the different dimensions of health status in patients with osteoarthritis (OA) [8]. The Western Ontario and McMaster OA index (WOMAC) and Lequesne index as a special measure of OA are mainly focused on the impact of the physical function, physical symptoms, and disease, and it cannot reflect other aspects of quality of life, such as psychological and social. The Medical Outcomes Study 36-Item Short-Form Health Survey (SF-36) and EuroQol five-dimensional (EQ-5D) have been widely used in studies of OA patients but are less sensitive to measure the results of specific disease intervention [9]. Most of KBD patients cannot understand the mean of some items in these questionnaires, because those patients come from remote mountainous areas and had low level of education and low understanding ability. Then, our team developed a new, simple, practical, 28-item instrument KBD Quality of Life (KBDQOL) that can capture KBD characteristics, satisfy the psychometric properties required for use in clinical trials and observational studies, and is easy to understand. However, in clinical practice, this new questionnaire has not been applied to evaluate the disease-specific quality of life in KBD patients before and after TKA surgery.

In this study, we tried to retrospectively analyze the clinical outcomes of 22 KBD patients who suffered severe OA in knee joint and underwent TKA surgery (25 knees) and to examine the quality of life in KBD patients before and after TKA surgery by a new first KBD-specific quality of life instrument of KBDQOL.

Methods

Patients

A total of 22 subjects (25 knees), who suffered end-stage KBD and severe knee pain, were retrospectively enrolled in this study between January 2007 and January 2016. All these

subjects underwent operation of primary TKA and were recruited from the Department of Orthopedics, Shaanxi Provincial People's Hospital, Xi'an, China.

The clinical diagnosis of KBD

KBD was diagnosed according to the clinical criteria [10].

Surgical technique

The same senior surgeon performed all operations. All the operations were performed through a standard midline incision and a medial parapatellar approach with patellar eversion in knee joint and using a cemented posterostabilized total knee prostheses (Chunli, Beijing, China).

Rehabilitation program for knee function

Passive knee joint exercises and full weight-bearing walking were started at two days post-surgery, when the drain was removed, and usually, most patients could passively achieve 90° flexion at this time. The knee active flexion was greater than 100° on the 14th post-operative day, when the patients could be discharged.

Outcome measures

The clinical evaluation and the radiological assessment were performed, and the data were collected from each patient at pre-operation, two weeks, one month, three months, six months, one year, two years, and three years after surgery. Subsequently, the patients were followed up once a year. Two independent investigators, who were not involved in the surgery, measured the pre-operative and post-operative assessments and to reduce observation bias.

Knee pain was measured on a visual analogue scale (VAS). Knee Society Clinical Rating System Score (KSS) was assessed as a functional evaluation.

KBDQOL was used to evaluate the disease-specific quality of life in KBD patients before and after surgery. The KBDQOL questionnaire demonstrated evidence of content validity, internal consistency, reliability, and construct validity, and it provides an objective tool for assessing quality of life of KBD [8].

The KBDQOL questionnaire is used to assess the patients' condition in the last month. KBDQOL instrument have 28 items and six domains, and its brief content is the following: physical function (seven items), activity limitation (five items), support of society (four items), economics (three items), mental health (five items), and general health (four items) [8]. Each item has five options, and all items have a score of 1 to 5. We calculate the average score of each domain as follows: the average score of physical function = $(Q1.1 + Q1.2 + Q1.3 + Q2.1 + Q2.2 + Q2.3 + Q3.4) / 7$, the average score of activity

limitation = $(Q1.4 + Q1.5 + Q1.6 + Q1.7 + Q1.8) / 5$, the average score of support of society = $(Q4.2 + Q5.5 + Q5.6 + Q5.8) / 4$, the average score of economics = $(Q6.1 + Q6.2 + Q6.3) / 3$, the average score of mental health = $(Q4.1 + Q4.4 + Q4.5 + Q4.6 + Q5.4) / 5$, and the average score of general health = $(Q7.1 + Q7.2 + Q7.3 + Q7.4) / 4$. We compared the average score of each domain in pre-operation with that in post-operation, and 5 is the best average score. For the detailed instruction of KBDQOL, see Table 1 and reference [8].

Statistical analysis

Demographic characteristics and clinical features were expressed as means with standard deviations (SD) or as absolute numbers. Categorical variables were analyzed by chi-

square test (χ^2). Continuous variables with normal distribution were analyzed by *t* test. Comparing between pre-operation and post-operation, VAS score, KSS score, knee joint activity, and varus/valgus deformities of the joints were analyzed by a paired samples *t* test. A two-sided *P* value of less than 0.05 was considered statistically significant.

Results

Baseline characteristics of the study population

The study population consisting of seven males and 15 females (25 knees), with a mean age of 57.55 ± 7.49 years, were diagnosed with KBD and underwent TKA. The detailed data

Table 1 The KBDQOL questionnaire

Abbreviated item content of KBDQOL (in the past 1 month)		Alternative answer and score				
		1	2	3	4	5
1	Going up or down one step of stairs (Q1.1)	Absolutely can't	Very difficult	Medium difficulty	A little difficult	No difficulty
2	Kneeling down (Q1.2)	Absolutely can't	Very difficult	Medium difficulty	A little difficult	No difficulty
3	Bending down (Q1.3)	Absolutely can't	Very difficult	Medium difficulty	A little difficult	No difficulty
4	Walking 1 km (Q1.4)	Absolutely can't	Very difficult	Medium difficulty	A little difficult	No difficulty
5	Walking 100 m (Q1.5)	Absolutely can't	Very difficult	Medium difficulty	A little difficult	No difficulty
6	Dressing yourself (Q1.6)	Absolutely can't	Very difficult	Medium difficulty	A little difficult	No difficulty
7	Doing heavy labor such as farm work (Q1.7)	Absolutely can't	Very difficult	Medium difficulty	A little difficult	No difficulty
8	Doing light labor such as cooking (Q1.8)	Absolutely can't	Very difficult	Medium difficulty	A little difficult	No difficulty
9	Pain in joints (Q2.1)	Every day	21d-29d	11d-20d	1d-10d	0d
10	Duration of taking pain killer in days (Q2.2)	Every day	21d-29d	11d-20d	1d-10d	0d
11	Morning stiffness (Q2.3)	Every day	21d-29d	11d-20d	1d-10d	0d
12	Frequency of sleeplessness (Q3.4)	Every day	21d-29d	11d-20d	1d-10d	0d
13	Feel happy (Q4.1)	Always	Often	Sometimes	Occasionally	No
14	Feel contribution to family duty (Q4.2)	Always	Often	Sometimes	Occasionally	No
15	Feel yourself is a burden to others (Q4.4)	Always	Often	Sometimes	Occasionally	No
16	Feel blue mood (Q4.5)	Always	Often	Sometimes	Occasionally	No
17	Feel embarrassed about bodily appearance Q4.6	Always	Often	Sometimes	Occasionally	No
18	Feel that no one take care of you (Q5.4)	Always	Often	Sometimes	Occasionally	No
19	Feel supported by your family (Q5.5)	Always	Often	Sometimes	Occasionally	No
20	Hang out, chat with neighbors (Q5.6)	Always	Often	Sometimes	Occasionally	No
21	Have someone help you when you need (Q5.8)	Always	Often	Sometimes	Occasionally	No
22	Economy difficult Q6.1	Always	Often	Sometimes	Occasionally	No
23	Borrow money Q6.2	Always	Often	Sometimes	Occasionally	No
24	Can't afford treating disease Q6.3	Always	Often	Sometimes	Occasionally	No
25	In general, how about your health? (Q7.1)	Very poor	Poor	Just so so	Good	Very good
26	Compared to the same age and gender people, how about your health? (Q7.2)	Much worse	Worse	Same	A little better	Much better
27	Compared to 1 month ago, how about your health? (Q7.3)	Much worse	Worse	Same	A little better	Much better
28	In general, how satisfied are you with your quality of life (Q7.4)	Very dissatisfied	Not satisfied	Just so so	Satisfaction	Very satisfied

KBDQOL KBD Quality of Life, Q question

of patients' demographic characteristics and clinical features were listed in Table 2. There were no serious complications such as periprosthetic joint infection, deep vein thrombosis, pulmonary thromboembolism, common peroneal nerve injury, and fracture that occurred in patients. All prosthesis components were well fixed and no loosening was found in patients (Figs. 1 and 2).

VAS score levels in pre-operation and post-operation

Baseline VAS score levels in these patients were observed in pre-operation, and the mean VAS scores in all patients were 6.79 ± 1.40 . The levels of VAS score were significantly decreased on two weeks (3.71 ± 1.13) and three months after TKA (2.27 ± 1.17). The VAS score continues to decline within six months after TKA (2.0 ± 0.89) and maintain a low level on one year (1.80 ± 0.72), two years (1.64 ± 0.51), and three years (1.86 ± 0.59) after TKA. The changes of VAS score levels on a longitudinal basis in every patient were summarized in Fig. 3.

Table 2 Demographic characteristics and clinical features of the patients

Demographic characteristics and clinical features	Patients
Gender, <i>n</i> (%)	
Male	7 (31.82)
Female	15 (68.18)
Age (years)	57.55 ± 7.49
Side, <i>n</i> (%)	
Right	11 (44.0)
Left	14 (56.0)
Body mass index, kg/m ²	23.32 ± 2.05
Varus/valgus deformities	
Varus, <i>n</i> (%)	21(84)
Valgus, <i>n</i> (%)	4(16)
Grade of KBD, <i>n</i> (%)	
Grade I	13 (59.09)
Grade II	6 (27.27)
Grade III	3(13.64)
Length of hospitalization, days	18.3 ± 3.4
Drainage volume, mL	432.0 ± 202.4
Receiving a haemostatic regimen (12 patients)	255.0 ± 80.4
No haemostasis regimen (10 patients) ^a	595.4 ± 125.6
Blood transfusion, milliliter	384.0 ± 336.3
Receiving a haemostatic regimen (12 patients)	100 ± 180.9
No haemostasis regimen (10 patients) ^a	646.2 ± 202.5
Surgery length, h	147.76 ± 56.13
Duration of follow-up, months	44.7 ± 30.3

^a Receiving a haemostatic regimen: tranexamic acid was administered intravenously 10 min before the operation started. No haemostasis regimen (10 patients) compared with receiving a haemostatic regimen (12 patients), $P < 0.001$



Fig. 1 A 45-year-old man who suffered from end-stage KBD and severe knee pain and underwent the operation of primary TKA. This case was introduced in our previous article [11] and needs to be supplemented in this article. This patient was characterized by a small pre-operative femoral condyle and tibial plateau (a). Conventional prosthesis cannot be used, thus the prosthesis need customization. The patient accepted right knee TKA (b). The post-operative position of the prosthesis was good. There was no loosening and the limb alignments had been improved

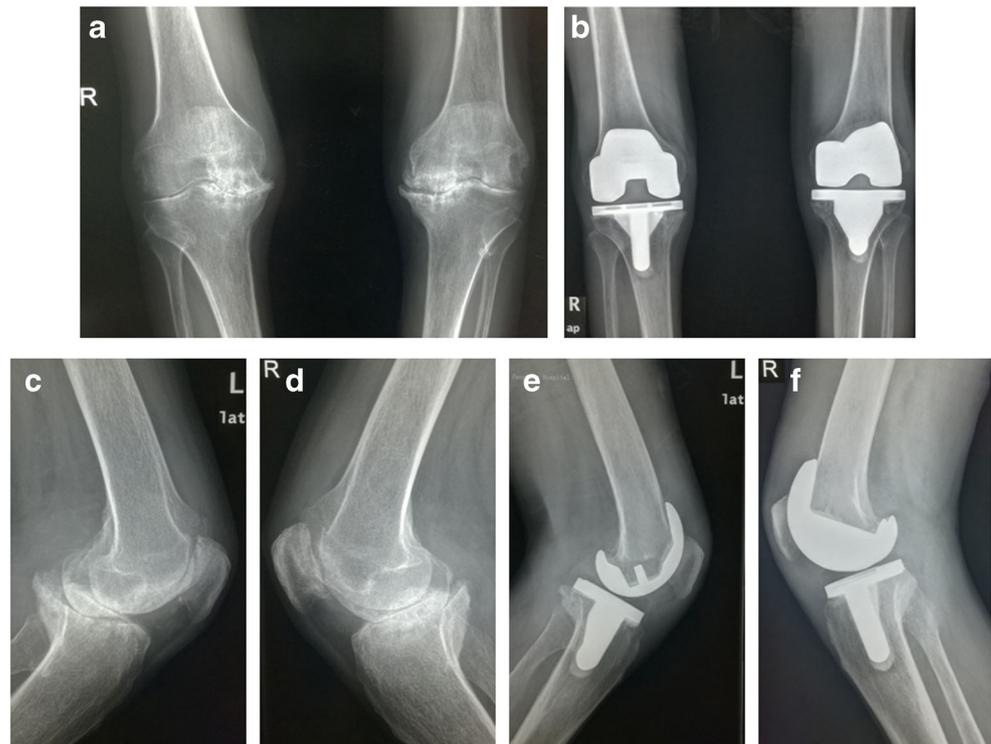
KSS score levels in pre-operation and post-operation

The patients' knee function was assessed by KSS (Fig. 4). The results highlighted that the level of KNEE score on 1 year after TKA (83.40 ± 10.89) was significantly higher than that in pre-operation (51.36 ± 24.32 , $P < 0.001$). At the same result, the level of FUNCTION score at one year after TKA (78.60 ± 10.95) was also higher than that in pre-operation (52.40 ± 22.64 , $P < 0.001$). There were no statistical differences in KNEE score between one and three years (91.5 ± 6.35) after TKA, ($P = 0.104$), and the levels of FUNCTION score at three years (89.17 ± 7.93) after TKA were significantly higher than those at one year after TKA ($P = 0.002$).

Varus/valgus deformities and range of motion of the knee in pre-operation and post-operation

The changes of the range of motion and degree of varus/valgus in patients' knee joints were evaluated in pre-operation and post-operation (Table 3). We only compared

Fig. 2 A 59-year-old man who suffered from end-stage KBD and severe knee pain and underwent the primary TKA operation. This patient was characterized by medial tibia plate pre-operative lip-like bone abrasion and severe joint narrowing (a, c, and d). Five years after surgery, all prosthesis components were well fixed and no loosening was found in this patient (b, e, and f)



the data results in post-operation with that in one year after TKA. The extension in pre-operation ($12.08^\circ \pm 11.30^\circ$) was significantly higher than that in one year after TKA ($3.04^\circ \pm 2.79^\circ$, $P < 0.001$). The flexion and range of motion in one year after TKA ($113.60^\circ \pm 14.55^\circ$ and $110.56^\circ \pm 15.58^\circ$, respectively) were significantly increased than that in pre-operation ($100.60^\circ \pm 22.97^\circ$ and $88.52^\circ \pm 32.11^\circ$, respectively, $P < 0.001$). The degree of varus or valgus in one year after TKA

($1.71^\circ \pm 2.31^\circ$ and $3.75^\circ \pm 2.5^\circ$, respectively) was significantly lower than that in pre-operation ($10.00^\circ \pm 9.08^\circ$ and $18.00^\circ \pm 5.72^\circ$, respectively, $P < 0.001$ and $P = 0.007$).

KBDQOL questionnaire in pre-operation and post-operation

The disease-specific quality of life in KBD patients was further assessed by KBDQOL questionnaire before and after surgery. The changes of the average score of each domain on a longitudinal basis in every patient were summarized in Fig. 5a–c, g–i. We found that the average score of physical function (4.33 ± 0.55), activity limitation (4.60 ± 0.55), support of society (3.66 ± 0.43), mental health (3.94 ± 0.46), and general health (3.27 ± 0.58) in one year after TKA was significantly higher than that in pre-operation (2.57 ± 0.76 , 4.23 ± 0.61 , 3.22 ± 0.47 , 2.65 ± 0.74 , and 1.92 ± 0.44 , respectively, $P < 0.001$, Fig. 5d–f, k, l), except for the average score of economics (pre-operation = 3.71 ± 1.08 , 1 year after TKA = 3.76 ± 1.00 , $P = 0.378$, Fig. 5j). Three years after TKA, only the average scores of physical function (4.73 ± 0.22) and mental health (4.28 ± 0.30) were significantly higher than that in one year after TKA ($P = 0.037$ and $P = 0.039$, respectively, Fig. 5d, k); however, the average scores of the other domains such as activity limitation (4.82 ± 0.22), support of society (3.95 ± 0.23), economics (3.83 ± 0.86), and general health (3.50 ± 0.47) were not statistically different with that in one year after TKA ($P > 0.05$, Fig. 5e, f, j, l).

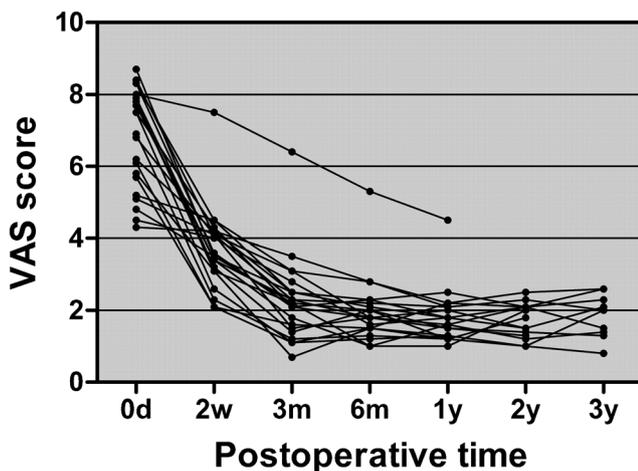


Fig. 3 Changes of VAS score levels on a longitudinal basis in every patient. High VAS score levels in these patients were observed in pre-operation, and the levels of VAS score were obviously decreased 2 weeks and 3 months after TKA. The VAS score levels were continually decreased 6 months after TKA and maintained a low level 1, 2, and 3 years after TKA. VAS visual analogue scale, TKA total knee arthroplasty

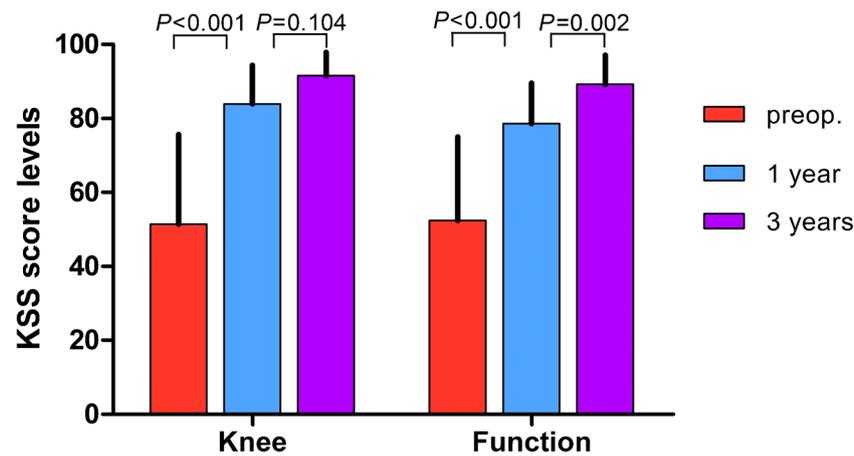


Fig. 4 KSS score levels in pre-operation and after TKA. The levels of KNEE score were increased 1 year after TKA (83.40 ± 10.89), compared with the pre-operative values (51.36 ± 24.32) and it maintained a higher level three years (91.5 ± 6.35) after TKA. The level of FUNCTION score

in pre-operation (52.40 ± 22.64), one year (78.60 ± 10.95), and three years (89.17 ± 7.93) after TKA increased accordingly. KSS Knee Society Clinical Rating System Score, *Preop.* Pre-operation, *TKA* total knee arthroplasty

Discussion

In this article, we respectively evaluated the clinical outcomes of TKA in adult KBD patients who suffered severe knee OA, and the results highlighted that TKA can reduce knee pain, improve knee function, correct the deformities of knees, and improve the quality life in KBD patients. Another important point is that this study was the first clinical evaluation, to our knowledge, to examine the quality of life in KBD patients before and after TKA surgery by a new KBD-specific quality of life instrument KBDQOL. Our data made KBDQOL

questionnaire a promising and elegant instrument for assessing the quality life in KBD patients who underwent TKA.

KBD is a chronic, deformable, endemic osteoarthropathy, and it is mainly prevalent in some areas of China, Russia, and North Korea [12]. The disease occurs in childhood and usually continues to progress, and it can cause joint pain, morning stiffness, and enlarged and shortened fingers, which are the primary symptoms of KBD. In the later stages of adulthood, KBD is characterized with deformed and enlarged joints and limited motion in the extremities [13]. The clinical symptoms mainly involve multi-joint pain, usually the shoulder, elbow, wrist, hip, knee, ankle, and interphalangeal joint [14].

The knee joint is often affected by the KBD disease with an estimated incidence of approximately 93%. Knee pain and functional limitations are the main factors which impact on adult KBD patients' daily life and work [14].

Without effective treatments, most of KBD patients will partly or completely lose their work and self-care abilities [15]. Therefore, relieving knee pain, reducing functional disability, and correcting the deformity are important aims for the treatment of adult KBD patients. The treatment of end-stage KBD remains a significant clinical challenge, and clinical data are lacking. Compared with OA, although KBD has a different aetiology, adult KBD lead to a similar pathological outcome that result in a secondary chronic osteoarthropathy, and the common clinical symptoms are joint pain, deformity, and severe dysfunction in the end-stage [16]. Therefore, the current KBD treatments are derived from the experiences in OA treatment.

As early as 2001, our research team tried to relieve knee pain in KBD patients by arthroscopic debridement [17–19]. Our results showed that the arthroscopic debridement cannot only reduce the short-term knee pain but also effectively maintain long-term effect in patients with KBD. However, the

Table 3 The changes of the range of motion and degree of varus/valgus in pre-operation and post-operation

	<i>n</i>	Mean (°)	SD (°)	<i>P</i> values
Extension				
Pre-op.	25	12.08	11.30	
1 year	25	3.04	2.79	<0.001
Flexion				
Pre-op.	25	100.60	22.97	
1 year	25	113.60	14.55	<0.001
Range of motion				
Pre-op.	25	88.52	32.11	
1 year	25	110.56	15.58	<0.001
Varus				
Pre-op.	21	10.00	9.08	
1 year	21	1.71	2.31	<0.001
Valgus				
Pre-op.	4	18.00	5.72	
1 year	4	3.75	2.50	0.007

P values, 1 year after TKA compared with pre-operation

Pre-op. pre-operation, *SD* standard deviations

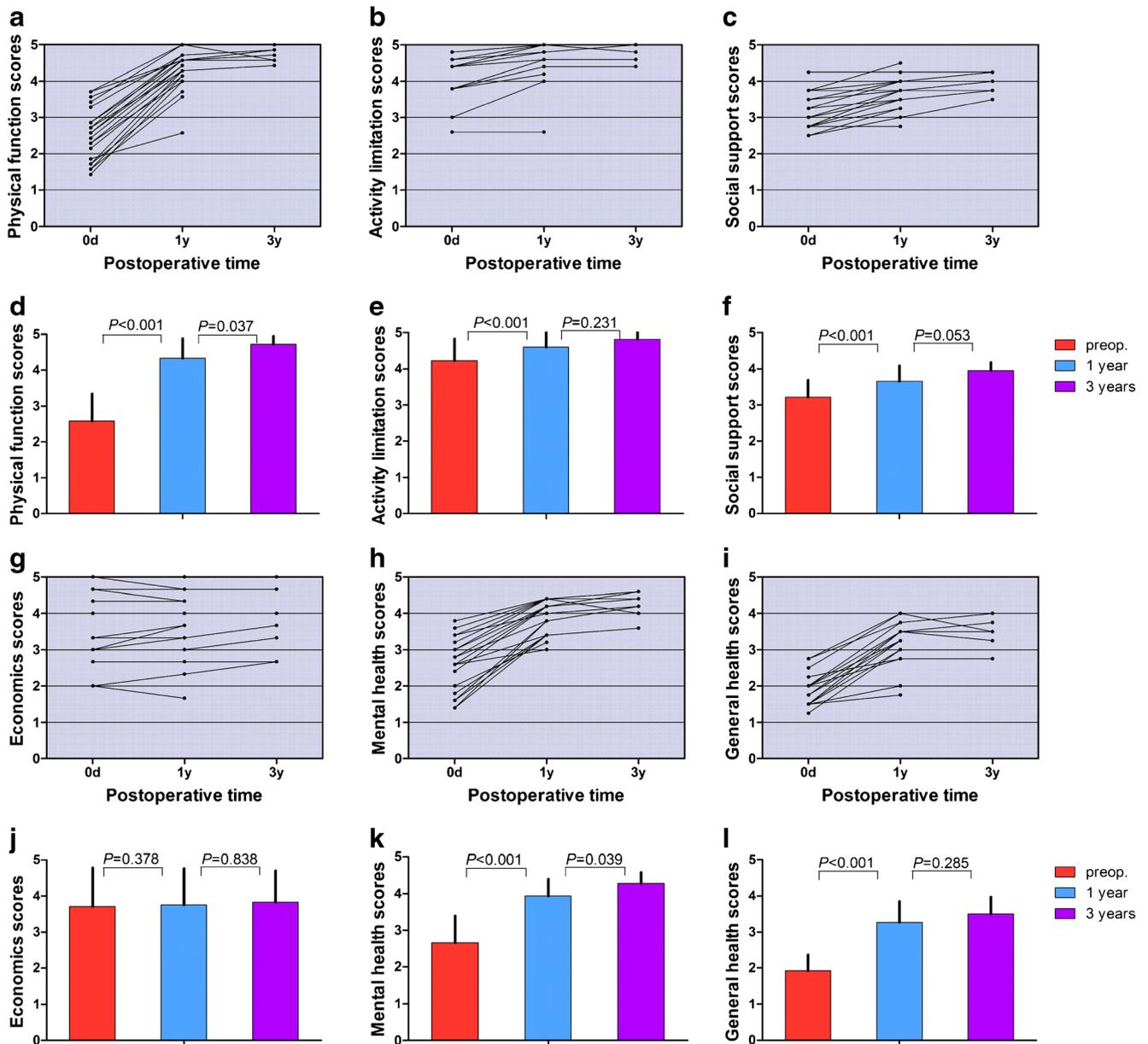


Fig. 5 The levels of KBDQOL questionnaire in pre-operation and after TKA. Changes of the average score level of each domain on a longitudinal basis in every patient were summarized in **a, b, c, g, h, and i**, and in most of patients, the average score level of each domain in pre-operation, 1 year, and 3 years after TKA increased accordingly. The average scores of physical function, activity limitation, support of society, mental health, and general health 1 year after TKA were significantly higher than that in pre-operation (**d, e, f, k, and l**), but there were no statistical differences

between the average score of economics in 1 year after TKA and those in pre-operation. Three years after TKA, the average scores of physical function and mental health were significantly higher than that in 1 year after TKA (**d, k**); however, the average scores of the other domains such as activity limitation, support of society, economics, and general health were not statistically different with that in 1 year after TKA (**e, f, j, l**). *KBDQOL* Kashin-Beck Disease Quality of Life, *Preop.* Pre-operation, *TKA* total knee arthroplasty

candidates for arthroscopic debridement were those with Kellgren-Lawrence grade I to III, and the patients with Kellgren-Lawrence grade IV, varus deformity, valgus deformity, and ligament injuries were excluded. Liu et al. [16] reported that osteotomy with impaction could correct the initial varus deformity and improve the function of knee joints in KBD patients, but it is not suitable for KBD patients with severe articular cartilage degeneration.

Therefore, the conventional methods cannot provide sufficient effect for patients who suffered advanced KBD, severe varus or valgus deformity, flexion deformity, and rotation mixed deformity in knee joints [7]. TKA is the most effective surgical procedure for improving physical function and relieving knee pain in patients who have severe OA of the knee [20]. We chose elderly KBD patients with pathologic lower limb mechanical axis, severe deformities, and severe articular

cartilage degeneration in knee joints to evaluate the efficacy of TKA for the treatment.

There were two reports that demonstrated better clinical and functional outcomes in patients with advanced KBD of the knee by a short-term follow-up. A previous study by Yang et al. [7] reported a favourable outcome in 23 KBD patients (27 knees) who underwent primary TKA. In this study, no radiographic evidence of loosening, sinking, or fracture in component positioning was found, and the average of hospital for special surgery knee score (HSS) after surgery was significantly higher than that before surgery. A recent study by Tang et al. [5] reported 15 KBD patients (18 knees) who received TKA treatment by average 32-month follow-up. In this study, the VAS score at the final follow-up (after surgery) was significantly decreased than that before surgery, and the HSS score at the final follow-up was significantly increased than that before surgery. In our research, the patients were followed up for an average of 44.7 ± 30.3 months. Similarly with the above two researches, our results showed that TKA had significantly relieved knee pain and improved the quality of life of KBD patients who had severe OA.

However, we found that the efficacy of TKA in KBD patients was worse than that in OA patients. There are several reasons. Firstly, most of the adult KBD patients suffered multiple joint damage [21], such as knees, ankles, elbows, hips, and hands, affecting health status and quality of life in patients after surgery. In our study of 19 patients, hip, ankle, elbow, or hand joint damage is involved. Secondly, most of the adult KBD patients suffered more severe muscle atrophy, genu valgum, flexion deformity, and bone defects, which make operation more difficult and causing insufficient efficacy. Thirdly, the postoperative rehabilitation exercise is an important contributing factor for excellent results [22].

The quality of life in KBD patients was measured by the KBDQOL questionnaire that is the first disease-specific quality of life instrument for KBD patients. In this study, we found that the average score of physical function, activity limitation, support of society, mental health, and general health in one year after TKA was significantly higher than that in pre-operation, and the average score of physical function and mental health in three years after TKA was significantly higher than that in one year after TKA. The results proved that TKA can improve the quality of life in KBD patients. However, the domains of economics were not sensitive to the change between pre-operation and post-operation. In fact, the patients' economic situation was not improved after surgery. Therefore, the average scores of economics were not statistically different between pre-operation and one year after TKA.

We started to use haemostatic drugs to control blood loss and reduce post-operative blood transfusion since 2014. The intra-operative haemostatic regimen was implemented on 12 patients who had lower levels of drainage volume and blood transfusion than other ten patients who did not received any haemostatic regimen.

Our study has potential limitations. Our results were obtained from a single centre. The sample size was relatively small. Because most of patients with KBD were located in economically backward and remote mountain areas, the patient's economic situation is poor. Although there were many patients with KBD, only a few patients can afford hospitalization costs and accepted the operation of TKA. Large scale multicentre studies need to be performed to further support our findings.

Conclusions

The results demonstrate that TKA can reduce knee pain, improve knee function, correct knee deformities, and make a satisfied quality of life in KBD patients. Our data made KBDQOL questionnaire a promising and elegant instrument for assessing the quality life in KBD patients with TKA.

Acknowledgments We thank Xue-Yuan Wu, Xiang-Hui Dong, and Ming Chen for their English editorial assistance.

Funding This study was supported by the Project of International Science and Technology Cooperation and Exchange of Shaanxi Province (2017KW-051) and the Shaanxi Provincial People's Hospital In-Hospital Incubation Fund (2017YX-04).

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflicts of interest.

Ethical approval The study protocol was approved by the Ethical Committee of Shaanxi Provincial People's Hospital (Approval number 20121202) and in strict accordance with the Helsinki Declaration. This was a retrospective and observational study with medical records; this was not an intervention study of human samples, thus consent to participate was not required. Our Ethics Committee also approved that consent to participate was not needed.

Informed consent A written informed consent was obtained from all individual participants included in the study before enrollment.

References

1. Hinsenkamp M (2001) Kashin-Beck disease. *Int Orthop* 25(3):133
2. Liu QY (2008) Reviewing focus of prevention and treatment of Kashin-Beck disease. *Chinese Journal of Endemiology* 27(5): 473–474
3. Luo R, Liu G, Liu W, Pei F, Zhou Z, Li J, Shen B, Kang P, Xie Q, Ma X (2011) Efficacy of celecoxib, meloxicam and paracetamol in elderly Kashin-Beck disease (KBD) patients. *Int Orthop* 35(9): 1409–1414
4. Yang P, Guo X, He X, Zang Q, Wang G, Xu P, Wang W (2016) The efficacy and safety of intra-articular injection of hyaluronic acid in the knee and physical therapy agents to treat Kashin-Beck disease: a prospective interventional study. *Exp Ther Med* 12(2):739–745

5. Tang X, Zhou ZK, Shen B, Kang PD, Yang J, Li J, Pei FX (2014) Total knee arthroplasty in elderly patients with severe Kashin-Beck disease of the knee. *Int Orthop* 38(4):753–759
6. Tang X, Zhu J, Zhou Z, Shen B, Kang P, Pei F, Li J (2016) Total hip replacement in adult patients with severe Kashin-Beck disease of the hip. *BMC Musculoskelet Disord* 17(1):289
7. Yang LG, Gao DL, Tu ZM, Dong S, Ren J, Xie JJ, Zhao X, Li R, Liu G (2012) Stabilized prosthesis after total knee arthroplasty for the adult Kashin-Beck disease with mixed deformities. *Chinese Journal Bone and Joint Surgery* 5(3):212–216
8. Fang H, Guo X, Farooq U, Xia C, Dong R (2012) Development and validation of a quality of life instrument for Kashin-Beck disease: an endemic osteoarthritis in China. *Osteoarthr Cartil* 20(7):630–637
9. Fang H, Guo X, Xia C, Wang D (2014) Introduction and guidance on the Kashin-Beck disease quality of life. *Chinese Journal of Endemiology* 33(4):446–449
10. Liu N (2010) Interpretation of diagnostic criteria for Kashin-Beck disease. *China Health Standard Management* 1(4):56–58
11. Ling M, Wu X, Chang Y, Dong X, Sun Z, Ling L, Wu S, Yang B (2017) Staged total knee arthroplasty for bilateral complex knee deformities from Kashin-Beck disease and skeletal dysplasia. *Knee* 24(3):692–698
12. Liu JT, Guo X, Ma WJ, Zhang YG, Xu P, Yao JF, Bai YD (2010) Mitochondrial function is altered in articular chondrocytes of an endemic osteoarthritis, Kashin-Beck disease. *Osteoarthr Cartil* 18(9):1218–1226
13. Huang Q, Zhou ZK, Ma J, Li Y, Yang X, Shen B, Yang J, Kang PD, Pei FX (2015) The arthropathic and functional impairment features of adult Kashin-Beck disease patients in Aba Tibetan area in China. *Osteoarthr Cartil* 23(4):601–606
14. Li Y, Zhou Z, Shen B, Yang J, Kang P, Yang X, Liu G, Pei F (2013) Clinical features of Kashin-Beck disease in adults younger than 50 years of age during a low incidence period: severe elbow and knee lesions. *Clin Rheumatol* 32(3):317–324
15. Farooq U, Guo X, Chuang LH, Fang H, Zhuang G, Xia C (2011) Measuring health-related quality of life in Kashin-Beck disease using EQ-5D. *Qual Life Res* 20(3):425–429
16. Liu FD, Wang ZL, Hinsenkamp M (1998) Osteotomy at the knee for advanced cases of Kashin-Beck disease. *Int Orthop* 22(2):87–91
17. Yi Z, Ling M, Luo Z (2006) Observation on treatment result of arthroscopic knee clearing and reconstructing operation in Kashin-Beck disease. *China Journal of Endoscopy* 12(9):937–939
18. Ling M, Huang X, Yi Z (2010) Evaluation of long term effects of arthroscopic knee debridement and reconstructing for treating osteoarthritis in patients with Kashin-Beck disease. *Chinese Journal of Endemiology* 29(5):559–561
19. Ling M, Sun Z, Yi Z, Chang Y, Liu S, Yang G, Tian X, Dong X (2015) Long-term efficacy of arthroscopic debridement on knee osteoarthritis in patients with Kashin-Beck disease. *Cell Biochem Biophys* 73(1):125–128
20. Dixon MC, Brown RR, Parsch D, Scott RD (2005) Modular fixed-bearing total knee arthroplasty with retention of the posterior cruciate ligament. A study of patients followed for a minimum of fifteen years. *J Bone Joint Surg Am* 87(3):598–603
21. Mathieu F, Begaux F, Suetens C, De Maertelaer V, Hinsenkamp M (2001) Anthropometry and clinical features of Kashin-Beck disease in central Tibet. *Int Orthop* 25(3):138–141
22. Brennan GP, Fritz JM, Houck LT, Hunter SJ (2015) Outpatient rehabilitation care process factors and clinical outcomes among patients discharged home following unilateral total knee arthroplasty. *J Arthroplast* 30(5):885–890