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Original Article

Haemophilic; Arthropathy: Assessment of Quality of Life After Total Knee Arthroplasty

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Abstract: The goal of this study was to examine how the known effects of total knee arthroplasty (TKA) on clinical outcome parameters translate into improved quality of life, as measured with validated condition-specific and generic questionnaires (Knee Society Score, WOMAC, SF-12, transition questions), addressing physical, mental and social health. Eleven patients (13 knees) undergoing TKA from 1986 to 1994, with the diagnosis of severe haemophilic arthropathy of the knee, were followed-up over a 4-year period on average. TKA was found to reduce the burden of disease to levels similar to patients with osteoarthritis undergoing hip arthroplasty. Clinical and functional improvement after TKA translated into a substantial and significant increase in quality of life and patient satisfaction, found in objective as well as in patient-perceived measures. However, the physical functional ability did not reach the same level as in the corresponding population not affected by haemophilia, due to residual symptoms and impairment of other joints.

Keywords: Arthritis; Haemophilia; Health status

Introduction

Progressive arthropathy is the consequence of recurrent haemarthrosis, which occurs in all patients suffering from severe or moderate haemophilia, particularly during childhood and adolescence, the knee being the most frequent site of clinical manifestations [1,2].

The pathogenesis of haemophilic arthropathy is complex and involves a vicious circle of chemical and biomechanical factors. Haemarthrosis induces chronic synovitis and pain; reflex inhibition and physical inactivity lead to atrophy of the quadriceps muscle, particularly of the vastus medialis. Recurrent soft-tissue bleeding is followed by fibrotic reorganisation. Proteolytic inflammation products cause cartilaginous destruction and intraosseous bleeding leads to subchondral bone erosion. The increased vascularity of the hyperplastic synovial membrane favours new bleeding episodes. The consequences are disabling pain, instability, flexion contracture and femoro-tibial subluxation [2–4].

The cornerstone of disease management is the substitution of the missing factor in controlled home therapy programmes. With prevention and early treatment of bleeding episodes, haemophilic arthropathy develops later in life and often occurs only in one joint [1,5].

With the development of factor concentrates in the early 1960s, surgical therapy of haemophilic arthropathy has become possible. Initially, static corrections and soft-tissue interventions were performed to reduce pain and bleeding incidence [6–12]. However, only with the introduction of joint replacement surgery [13–15] could functional limitations be treated effectively [16–18].

The goal of this study was to examine how the known effects of total knee arthroplasty (TKA) on range of motion, stability and pain translate into improved quality of life, as measured with validated condition-specific and generic questionnaires [19], addressing physical, mental and social health.

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Materials and Methods

Study Design and Patient Selection

We performed a retrospective cohort study using clinical examination, patient questionnaires and chart review.

Fourteen consecutive male patients undergoing TKA at our institution from October 1986 to December 1994, with the diagnosis of severe haemophilic arthropathy of the knee, were included in this study. At the time of the follow-up in August 1995, three had died of causes not connected with TKA, thus 11 patients (13 knees) could be included. The average age at time of surgery was 44 years, ranging from 23 to 68 years. The length of the follow-up was 4 years and 2 months on average and ranged from 8 months to 8 years and 10 months. All participants were patients for many years at the interdisciplinary consultation service for haemophiliacs, which is run by an orthopaedic surgeon, a rheumatologist, a haematologist and a physical therapist.

Intervention

Preoperative standard procedures included a physical examination, standing antero-posterior and lateral radiographic studies of the involved knee, and a careful haematological evaluation, with special regard to the exclusion of factor VIII / IX inhibitors. All patients were treated by a standard prophylactic antibiotic regimen with cephalosporin and by replacement therapy with factor VIII / IX concentrate to a preoperative level of 100 IU/dl [16].

All procedures were performed under neuroleptanalgesia in a conventional operating theatre with a vertical laminar downflow system. Electro-coagulation was used, tourniquet being applied in most cases. The medial parapatellar exposure was employed in all cases, the unconstrained PCA and Duracon total prostheses were implanted cementlessly by a single surgeon and total synovectomy was always performed [20]. Rehabilitation was planned and monitored by the above-mentioned interdisciplinary team.

Data Collection and Measurement Instruments

Pre- and perioperative data including patient characteristics, intervention specifics and complications were collected from chart review.

Standardised preoperative and follow-up clinical examinations were done by the orthopaedic surgeon (Knee Society Score [21]) and the physical therapist (muscle strength). The Knee Society Score includes seven items addressing pain, range of motion, stability, deformity, and, as functional outcome parameters, maximal walking distance, stair climbing and the need for walking assistance. Isometric flexion and extension strength of the knee was measured with a hand-held pull gauge [22] with a good inter- and intraobserver reliability (Pearson's correlation coefficient 0.94 each).

Patient-perceived health, as assessed by self-administered validated questionnaires, was examined at a follow-up visit in 1995. Generic and condition-specific questionnaires were filled out twice, once for the current status and once by remembering the preoperative situation.

General health was measured with the 12-item questionnaire SF-12 [23] providing a physical and mental health profile. The score ranged from 14 to 70, with 70 representing perfect health.

Condition-specific health was measured with the Western Ontario and McMaster Questionnaire (WOMAC) [24,25], consisting of 24 items addressing the three dimensions: stiffness, pain and physical function. Items were scored on a 0–10 numerical rating scale, dimension scores were calculated as the mean of non-missing items, if the missing items were equal or less than 10% of the total number of items. The dimension scores ranged from 0–10, with 10 representing worst pain, stiffness and physical function.

Preoperative co-morbidity [26,27] was measured with a short self-administered patient questionnaire that included conditions according to their frequency in general practice. Patients were asked whether they had one or more of the selected problems, if they received treatment for it, and whether or not they perceived limitations because of the problem.

Patient-perceived change between the pre- and postoperative situation was examined using transition questions addressing symptoms, physical and social functioning, activities of daily living and quality of life. The questions used Likert response scales with the categories: much worse, somewhat worse, the same, somewhat better, much better. Overall satisfaction with respect to the result of surgery was measured on a numerical rating scale with the anchors of completely dissatisfied and completely satisfied.

Non-parametric and parametric methods were used as appropriate for descriptive statistics and analysis of change.

Results

Patients and Intervention

Ten patients had severe haemophilia and one patient had moderate haemophilia. While the majority of patients had no or only mild co-morbid conditions, the two oldest patients (67 and 68 years of age) were suffering from well-controlled hypertension. Two patients underwent interventions of the lower extremity before TKA (arthrodesis of the opposite knee; arthrodesis of the ankle on the same side as the TKA).

At the time of operation, four patients had six joints affected, involving both the knees, the ankles and the elbows. A fifth patient had the above-mentioned joints

affected as well as a hip joint. Four patients had three or four joints affected and two patients had only their knees involved.

Preoperatively, four knees had a Kellgren and Lawrence score of IV and nine knees a score of III–IV [28].

Nine patients had undergone unilateral procedures and two patients bilateral procedures. The type of prosthesis was a PCA in seven and a Duracon in six knees. In one patient the arthroplasty was combined with a supracondylar osteotomy.

In six out of the 13 interventions, articular bleeding complicated the postoperative course requiring arthrocentesis (four cases), arthroscopic arthrocentesis (one case) or open haemostasis (one case) because of recurrent bleeding. Hospitalisation time ranged from 23 to 58 days with a mean of 35 days.

Radiographic follow-up showed no signs of loosening of the prosthesis components. No implant had to be revised or became infected.

Clinical Outcomes

Table 1 shows the clinical health outcomes. According to the Knee Society Score the overall result was excellent or good in seven and moderate in six knees. Three patients with only a moderate response had varus deformity or persistent flexion contractures and two patients had a relevant extension deficit (15° and 25°, respectively). The average range of motion increased from 63° to 101°. Nine knees were completely free of pain and four ached only slightly or occasionally, while before intervention two knees were quite painful and eight were constantly severely painful. Specifically, functional improvement was rated as good in one and excellent in 12 knees. No single item of the knee score showed a deterioration in any patient.

Of the five patients available for pre- and postoperative strength measurements, knee extension strength increased significantly, while no increase in knee flexion strength was recorded (Table 1).

Patient Perceived Outcomes

Overall, the intervention was rated as completely satisfying for 10 and quite satisfying for two knees. One patient was indifferent about the result. With the exception of this patient, there was generally much improvement perceived in most transition questions. Two patients reported no improvement in articular bleeding due to ongoing bleeding episodes in other joints.

Consistent with the transition questions (Table 2), there was a significant and important improvement of all WOMAC dimensions (Table 1), apart from the mentioned exception.

General health at follow-up was around the 50th percentile with respect to the mental dimension of the SF-12 (51; population range 35–62) but beneath the 25th percentile with respect to the physical dimension (42; 28–52). Of the three patients scoring beneath 30 was one with a previous arthrodesis of the ankle and the two oldest patients with moderate co-morbidity, of which one perceived a worse outcome as measured with the WOMAC.

Discussion

Patients with haemophilic arthropathy experience an important reduction of their health status. The burden of disease is comparable to patients with osteoarthritis (OA) undergoing hip arthroplasty. Pain and function, as measured with the WOMAC, are comparable to a cohort of OA patients reported by Laupacis et al. [29] (pain: 5.4 compared with 5.0 in OA; functional limitation: 5.5 compared with 5.7 in OA). However, different from patients with OA, patients with haemophilia seem to experience more severe stiffness (7.5 compared with 5.9 in OA). This stiffness may be due to the severe intra-articular and soft-tissue fibrosis, typical features of chronic haemophilic arthropathy. Compared with patients with OA of the knee requiring non-steroidal

Table 1. Change in patient outcomes

	Preoperative		Follow-up		Change			*t*-test	*p*-value	Effect size	Standardised response mean
	Mean	SD	Mean	SD	Mean	SD	Mean (%)				
WOMAC											
Pain	5.38	3.08	0.37	0.84	5.01	3.26	93	5.54	<.001	1.63	1.54
Stiffness	7.47	2.87	0.85	1.36	6.62	3.65	89	6.54	<.001	2.31	1.81
Function	5.54	2.09	0.96	1.22	4.58	2.30	83	7.18	<.001	2.19	1.99
The Knee Society Score											
Knee	17.23	15.40	75.69	11.76	58.46	19.99	339	10.54	<.001	3.80	2.92
Function	49.23	11.90	96.15	6.25	46.92	13.47	95	12.56	<.001	3.94	3.48
Strength (*n*=5)											
Flexion, kp	12.30	3.73	13.30	4.32	1.00	1.27	8	1.76	<0.1	0.27	0.79
Extension, kp	11.60	7.58	21.20	12.74	9.60	9.38	83	2.29	<.025	1.27	1.02

Table 2. Patient evaluation of change (transition questions for 13 surgical procedures)

	Much worse	Somewhat worse	No change	Somewhat better	Much better
In general				1 (8%)	12 (92%)
Pain			1 (8%)		12 (92%)
Stiffness (morning)			2 (15%)		11 (85%)
Mobility			1 (8%)	1 (8%)	11 (85%)
Stair climbing			2 (15%)		11 (85%)
Walking				1 (8%)	12 (92%)
Bending down			1 (8%)	3 (23%)	9 (69%)
Dressing and body care			2 (15%)	1 (8%)	10 (77%)
Housekeeping			1 (8%)	2 (15%)	10 (77%)
Social activities			1 (8%)	1 (8%)	11 (85%)
Work			1 (8%)	2 (15%)	10 (77%)
Mood		1 (8%)			12 (92%)
Life quality			1 (8%)		12 (92%)
Articular bleeding		1 (8%)	1 (8%)	2 (15%)	9 (69%)

antirheumatic treatment who reported WOMAC scores around 4.5 [30], the burden is substantially higher in haemophilia requiring joint arthroplasty.

Compared with routine TKA for degenerative arthritis [31], we found a high complication rate (in particular bleeding) in our patients. The complication rate in our study is comparable to other studies with haemophiliac patients [16–18].

TKA also reduces the burden of disease to levels similar to patients with OA undergoing hip arthroplasty (pain: 0.37 compared with 0.7; stiffness: 0.85 compared with 1.0; function 0.96 compared with 0.7 in OA, respectively) [29]. The percentage changes and responsiveness statistics are among the highest reported for interventions in medicine [32]. The substantial improvement in quality of life was found both in objective as well as in patient-perceived measures. Specifically, patients benefit in terms of social contacts and work ability, use of public transport, leisure and sport activities.

Nevertheless, the physical function ability does not reach the same level as in the corresponding population not affected by haemophilia: while at follow-up mental health, as measured with the SF-12, is on the 50th percentile, physical health is beneath the 25th percentile. It is important to note that muscle strength for both extension and flexion (both beneath the 5th percentile) is low. The remaining physical function disability may be due to the arthropathic affliction of other joints.

Our study is limited owing to the small sample size. However, because most patients with haemophilic arthropathy in a catchment area of around two million are seen at our institution, the results are likely to be representative. A second limitation is the retrospective assessment of patient-perceived outcomes. In a study with pancreas-kidney transplant recipients it has been found that patients rate their preoperative health worse in a retrospective than in a prospective assessment [33]. This may be explained by the fact that patients have adjusted well to their limitations before the intervention, but after the intervention put their prior health status in perspective of a considerably better health status. Assuming that this bias is also present in our population, the preoperative burden of disease as assessed in our study may be higher than in a study measuring preoperative burden prospectively. As a consequence, the prior burden of disease and the response to the treatment are likely to be overestimated. However, the agreement of the various approaches, particularly with the Knee Society Score, and the strikingly similar WOMAC profiles when compared with OA patients, supports the validity of the results.

While the WOMAC has not been specifically validated for patients with haemophiliac arthropathy, the WOMAC is now widely used in patients with impairments of the lower extremities that not only derive from primary OA. In particular, the WOMAC has been used in patients with, for example, avascular necrosis, rheumatoid arthritis and childhood hip disease [34].

In conclusion we found that clinical and functional improvement after TKA translates into a remarkable and significant increase in quality of life and patient satisfaction, despite considerable persistent impairment due to residual symptoms and impairment of other joints.

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