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Total Knee Arthroplasty in Patients With Juvenile Idiopathic Arthritis

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Abstract

Background Total knee arthroplasty (TKA) for juvenile idiopathic arthritis is rare but is nonetheless indicated for many patients with this disease. Few reports exist on the results of TKA in patients with juvenile idiopathic arthritis.

Questions/purposes It was sought to determine (1) survivorship and (2) functional outcomes of TKAs in patients with juvenile idiopathic arthritis.

Methods Results were combined from patients treated by experienced surgeons at five hospitals between 1979 and 2011. Two hundred nineteen patients (349 TKAs) were identified and contacted to survey their outcomes at a minimum followup of 2 years (mean, 12 ± 8 years; range, 2–33 years). The average age at surgery was 28.9 ± 9.7 years (range, 11–58 years). Data on revision surgery and ability to perform daily activities were collected.

Results The 10-year survivorship was 95%, decreasing to 82% by 20 years. At latest followup, 31 of 349 TKAs (8.9%) had been revised for either polyethylene failure or loosening (18 TKAs), infection (four), stiffness (three), periprosthetic fractures (two), bilateral amputation for vascular reasons (two), patellar resurfacing (one), and instability (one). Walking tolerance was unlimited in 49%, five to 10 blocks in 23%, and less than five blocks in 28%. Eleven percent could not manage stairs, and another 59%

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depended on railings. A cane was used by 12% and crutches by 7%; 12% were wheelchair-dependent.
Conclusions TKA survivorship in patients with juvenile idiopathic arthritis was inferior to that typically seen in younger patients with osteoarthritis or even rheumatoid arthritis confirming results of earlier studies with smaller patient numbers. This is especially disconcerting because younger patients require better durability of their TKAs.
Level of Evidence Level IV, therapeutic study. See Guidelines for Authors for a complete description of levels of evidence.

Introduction

TKA is a successful procedure for treatment of end-stage arthritis. National TKA registries report survival rates in patients with osteoarthritis of nearly 95% at 10 years and just below 90% at 20 years [11, 16]. Survival rates of TKA in patients with rheumatoid arthritis are slightly less favorable with rates of 93% at 10 years and 85% at 20 years [11, 18].

Most TKAs are performed in elderly patients. TKAs in very young patients are more rare because fewer young patients have painful knee problems requiring replacement. Indications in young patients include hemophilia, juvenile idiopathic arthritis, posttraumatic arthritis, and skeletal dysplasia. Most of these disorders cause early growth plate closure with significant bony deformities that make surgery technically challenging. Many patients with juvenile idiopathic arthritis are prescribed immunosuppressive medications or biological agents that may make them prone to infection. Patients with hemophilia also have higher infection rates and may have bony abnormalities and joint contractures. Posttraumatic patients with arthritis may resume high-impact activities that can lead to early failure. Thus, clinical results in young patients are inferior. For example, the Swedish registry showed revision rates approximately three times higher in younger (< 65 years) as compared with older (> 75 years) patients [11].

Juvenile idiopathic arthritis can be disabling. It commonly occurs in children between 7 and 12 years of age. Besides fever and rash, the disease is associated with joint involvement in most cases. The incidence of juvenile idiopathic arthritis is approximately six per 10,000 children [1], and it affects an estimated 294,000 children in the United States alone [8]. It is the leading cause of childhood disability with the knee involved in approximately two-thirds of patients, many with functional deficits [1].

As a result of premature growth plate closure in juvenile idiopathic arthritis, tibias and femurs often present with a trumpet-shaped deformity with abnormal AP and mediolateral dimensions compared with normal anatomy (Fig. 1). Patients with this disease often have valgus alignment with a flexion contracture and external tibial rotation. Bone quality is also frequently compromised, thus reducing the ability to secure durable implant fixation. As a result of the altered anatomy and small size of their knees, patients with juvenile idiopathic arthritis often cannot be treated with standard implants, so custom devices may be required (Fig. 2). Patients with juvenile idiopathic arthritis also frequently present with multiple joint involvement and significant functional limitations.

Only a few studies (Table 1) reported clinical results for TKA in patients with juvenile idiopathic arthritis with small numbers of cases at short-term followup [2, 5, 6, 12, 15]. We therefore sought to determine (1) survivorship and (2) functional outcomes of TKAs in patients with juvenile idiopathic arthritis.

on file with the publication and can be viewed on request.
Each author certifies that his or her institution approved the human protocol for this investigation, that all investigations were conducted in conformity with ethical principles of research, and that informed consent for participation in the study was obtained.
Data were collected at the University of California at San Francisco Medical Center, San Francisco, CA, USA; University Hospital Leuven, Pellenberg, Belgium; Stanford Medicine Outpatient Center, Redwood City, CA, USA; Brigham and Women's Hospital Physician Organization, Boston, MA, USA; and the Hospital for Special Surgery, New York, NY, USA. Data analysis was conducted at the Hospital for Special Surgery, New York, NY, USA.

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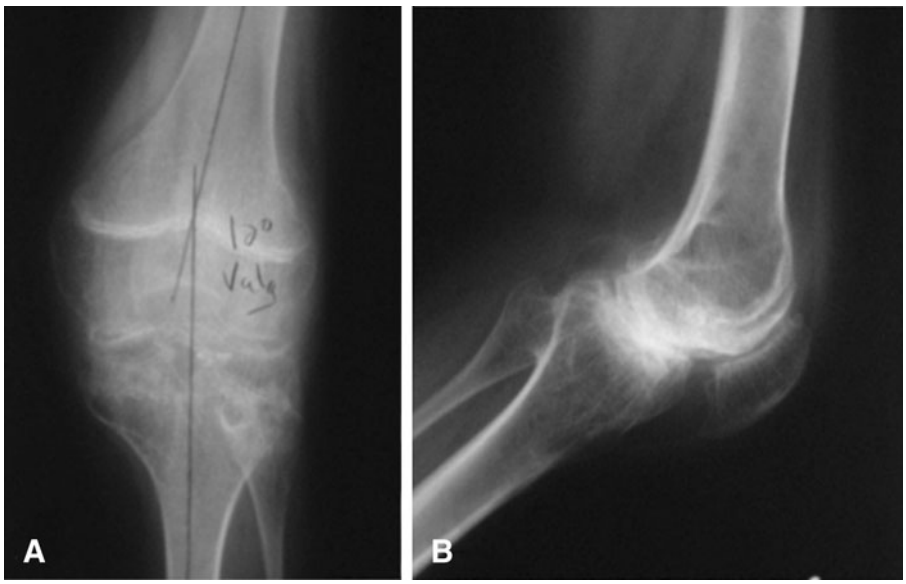

Fig. 1A–B Typical juvenile idiopathic arthritis deformity with a trumpet-shaped distal femur and proximal tibia, valgus deformity with flexion contracture, and patella baja in radiographs in two planes (**A**–**B**).

Patients and Methods

To accumulate sufficient numbers of TKAs for juvenile idiopathic arthritis (Fig. 3), an international multicenter study was initiated to combine results from patients treated by experienced arthroplasty surgeons at five hospitals: Hospital for Special Surgery, New York, NY, USA; Brigham and Women's Hospital, Boston, MA, USA; Stanford University Medical Center, Stanford, CA, USA; University Hospitals, Leuven, Belgium; and the University of California, San Francisco Medical Center, San Francisco, CA, USA. This institutional review board-approved study consisted of a chart review and patient followup by telephone. We identified 219 patients with 349 TKAs performed between the years 1979 and 2011. Inpatient charts were reviewed for preoperative diagnosis, surgical date, medication history, other joint involvement, and complications. Consenting patients were administered a questionnaire that was specifically designed for this study to survey their outcomes. Patients were surveyed either at recent followup or by telephone. Information on diagnosis, date, and side of surgery was confirmed and the need for additional surgery was obtained. Information on pain levels (0–10) with activity was obtained. Patients were asked if they had a sense of knee instability ("yes" or "no"). The walking tolerance was documented (unlimited, five to 10 blocks, less than five blocks) as well as the ability to ascend and descend stairs (without use of railings, with use of railings, unable), and the necessity to regularly use assistive devices (none, cane, crutches, walker, wheelchair). Patients were also asked if and which other bone or joint involvement limited their activity.

The average age at surgery was 28.9 ± 9.7 years (range, 11–58 years) with a minimum followup of 2 years (mean, 11.8 ± 7.8 years; range, 2–33 years). Fourteen patients (with 19 TKAs) had died, one bilateral patient refused participation, and nine patients (4.1%), with 13 TKAs (3.7%), were lost to followup.

Complete preoperative data were available on 208 patients (95%), and 181 (87%) had polyarticular disease, whereas 131 (63%) had undergone previous THA. One hundred sixty-two patients (78%) were on disease-modifying antirheumatic drugs before their surgery with methotrexate being the most common at 40% (83 patients). One hundred eight patients (52%) were on steroids, but 44 patients (21%) were only on nonsteroidal antiinflammatory drugs before TKA. One hundred ninety-eight of 219 patients (90%) had bilateral knee disease, and 130 (66%) of those patients had both knees replaced during the course of the study. Of these, more than half had undergone simultaneous bilateral TKA.

All but five of the TKAs were cemented; 85 (24%) were cruciate-retaining implant designs, 195 (56%) were posterior-stabilized designs, whereas 65 (19%) TKAs had constrained articulations. Hinged implants were used in four (1%) of the cases. Three hundred thirty-two (95%) knees underwent patellar resurfacing. Patient-specific custom-designed TKAs were used in 65 (19%) cases (Fig. 2). All of the custom implants were performed at the Hospital for Special Surgery in New York.

Descriptive analyses were performed and Kaplan-Meier survival analysis was also conducted using Microsoft Excel (Microsoft Corporation, Seattle, WA, USA) and IBM SPSS Statistics 21 (PASW 21; SPSS Inc, Chicago, IL, USA).

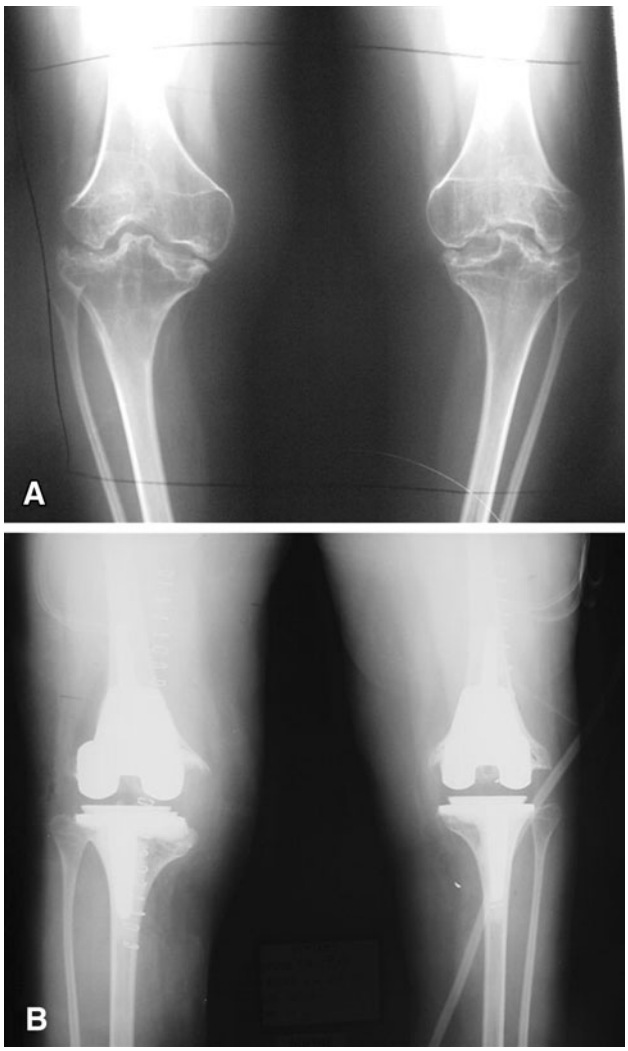

Fig. 2A–B The AP radiographs of this patient with juvenile idiopathic arthritis show bilateral marked joint destruction (**A**). Custom total knees were used as a result of patient size and offset of the tibial stem (**B**).

Results

Survivorship

The 10-year survivorship was 95%, which had decreased to 81.6% at 20 years (Fig. 4). There were differences in survivorship across the participating centers (Table 2). At latest followup, 31 of 349 TKAs (8.9%) had been revised. Polyethylene failure and its sequelae, loosening and osteolysis, were the main reasons for revision (n = 18 [58%]). Seven of the 20 knees revised for polyethylene wear and loosening had revision of their polyethylene component only, allowing for retention of their femur and tibial tray. Four revisions were done for infection (1.2% of all cases), which is surprisingly low given that 78% of patients were on immunosuppressive medications. Of the other revisions, three were performed for stiffness, two for periprosthetic fractures, two for vascular reasons that resulted in bilateral amputation, one for patellar resurfacing, and one for midflexion instability.

The 10-year survivorship based on implant constraint was 95% for the constrained knees, 92.6% for the posterior-stabilized knees, and 98.6% for the cruciate-retaining knees (Table 3). The 20-year survivorship rates were 92% for the constrained knees, 88.2% for the posterior-stabilized knees, and 69.2% for the cruciate-retaining knees. The custom knees had a 10-year survivorship of 91.7% as compared with

Table 1. Overview of literature with focus on TKA in patients with juvenile idiopathic arthritis

Lead author	Journal	Number of patients and knees	Revision rate (%)	Average followup (years)
Sarokhan [17]	JBJS Am, 1983	28/48	12.5	5
Carmichael [4]	CORR®, 1986	13/25	0	5.1
Parvizi [15]	JBJS Am, 2003	13/25	16	10
Bellemans [2]	Acta Orthop Belg, 1997	15/25	8	7
Malviya [12]	Knee, 2010	20/34	41.5	20
Jolles [9]	CORR®, 2008	14/22	0	8
Palmer [14]	JBJS Am, 2005	8/15	20	15.5
Thomas [19]	CORR®, 2005	10/17	5.9	6
Boublik [3]	CORR®, 1993	14/22	4.5	3.9

JBJS Am = *Journal of Bone and Joint Surgery, American*; CORR® = *Clinical Orthopaedics and Related Research*; Acta Orthop Belg = *Acta Orthopaedica Belgica*.

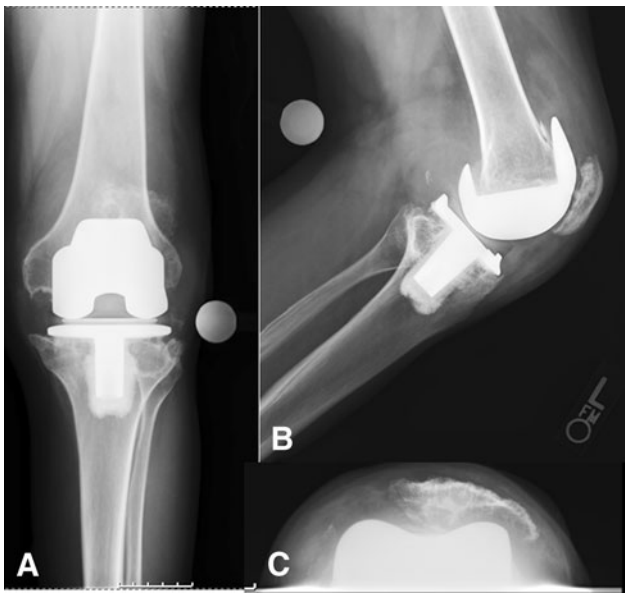

Fig. 3A–C Radiographs in three planes (**A**–**C**) of a left knee 25 years after TKA in a male patient, 17 years old at the time of the index procedure, are shown. This patient with multiple joint involvement was revision-free at latest followup and walks daily > 1 mile without walking aids.

the standard knee rate of 95.8%, but by 20 years, the survivorship rate of the custom implants had decreased to 89.7%, whereas the conventional knee rate was 77.8%. Differences in patterns of constraint use may have contributed to the observed differences in implant survivorship among the participating centers (Table 2). Other factors that may have influenced the variations in survivorship include severity of disease and functional levels at the time of surgery.

Functional Outcomes

Of the 196 patients who responded to the survey, walking tolerance was unlimited in 96 (49%), five to 10 blocks in 45 (23%), and less than five blocks in 55 (28%) at latest followup. Twenty-two of the patients (11%) could not manage stairs, and another 116 (59%) depended on railings when managing stairs. A cane was used by 24 (12%) during ambulation, and crutches were used by 14 (7%). A total of 24 (12%) of the patients were wheelchair-dependent. The average pain level (0–10) with activity was 1.1 ± 2.3 (range, 0–10). Eighteen (9%) of the patients reported a sense of instability of the operated knee.

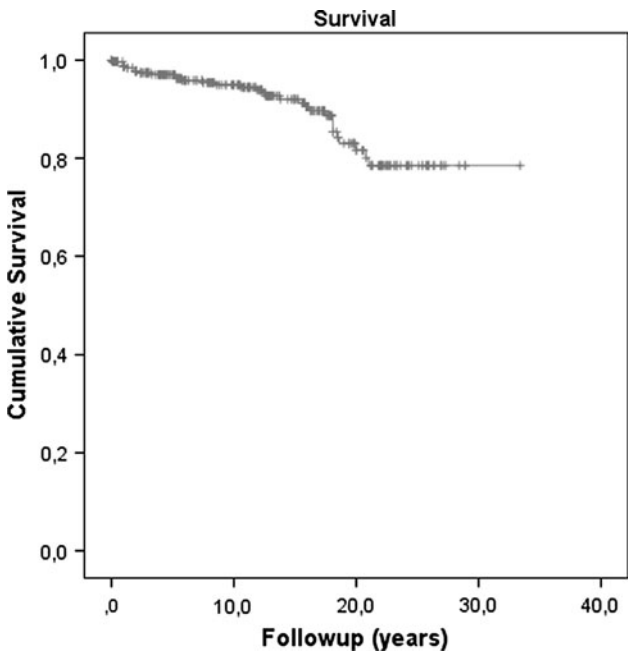

Fig. 4 Kaplan-Meier analysis revealed a 10-year survival rate of 95% and a 20-year survival of 81.6% of TKA in patients with juvenile idiopathic arthritis. Numbers of included TKAs in patients with juvenile idiopathic arthritis per year are shown.

Discussion

There is a paucity of adequately sized clinical long-term studies for TKA in patients with juvenile idiopathic arthritis [2, 5, 6, 12, 15]. With this international multicenter approach it was sought to determine (1) survivorship and (2) functional outcomes of TKAs in a large patient cohort with this disease. Many of the patients in this series experienced good or excellent function into the third and fourth decades after TKA despite their young age and often severe patterns of inflammatory arthritis. However, the survivorship was lower than those reported in the literature for older patients undergoing TKA [11, 16] but was comparable to that reported in other studies of similarly young patients [2, 12]. This is especially disconcerting because younger patients require greater longevity and better durability from their TKAs.

Although this study benefited from its multicenter design, like all studies, it had a number of limitations. Important differences regarding indications for TKA and their revision may exist among the participating surgeons as well as differences in techniques and implants and the manner of data collection over more than three decades. The learning curve of the surgeons is also hard to quantify. The influence of more aggressive medical therapy is difficult to ascertain, but our experience is that recent patients with juvenile inflammatory arthritis are presenting with more pauciarticular disease. In addition, although the total number of TKAs performed for juvenile inflammatory arthritis may be decreasing, the numbers of those performed at these institutions have remained relatively constant,

Table 2. Revision rates and survival by center

Center	Number of knees	Implant constraint	Revision rate (%)	Survival at 10 and 20 years (%) with SEs	
A	74	CR: 71, PS: 3	18.9	98.5 (97.0–100)	71.1 (64.2–78.0)
B	171	CR: 3, CCK: 59, PS: 106, RTH: 3	6.4	93.2 (91.1–95.3)	92.1 (89.8–94.4)
C	27	CR: 6, CCK: 2, PS: 18, RTH: 1	7.4	96.2 (92.4–100)	91.3 (85.4–97.2)
D	32	CR: 5, CCK: 3, PS: 24	0	100	100
E	45	CCK: 1, PS: 44	8.9	90.0 (83.0–97.0)	45.0 (13.0–77.0)
All centers	349	CR: 85, CCK: 65, PS: 195, RTH: 4	8.9	95.0 (93.7–96.3)	81.6 (78.0–85.2)

CR = cruciate-retaining; PS = posterior-stabilized; CCK = constrained; RTH = rotating hinge.

Table 3. Revision rates and survival by constraint

Constraint	Number of knees	Mean followup (years ± SD)	Revision rate (%)	Survival at 10 and 20 years (%) with SEs	
CCK	36	9.9 ± 5.7	8.3 (3)	94.4 (90.5–98.3)	94.4 (90.5–98.3)
CCK custom	29	15.9 ± 6.8	6.9 (2)	96.0 (92.1–99.9)	90.4 (83.8–97.0)
All CCK	65	12.4 ± 6.8	7.7 (5)	95.0 (92.2–97.8)	92.0 (87.9–96.1)
CR	85	14.8 ± 7.1	16.5 (14)	98.6 (95.3–99.9)	69.2 (62.0–76.4)
PS	160	9.1 ± 7.9	5.0 (8)	93.9 (91.4–96.4)	87.2 (81.5–92.9)
PS custom	35	15.9 ± 7.4	11.4 (4)	88.3 (82.8–93.8)	88.3 (82.8–93.8)
All PS	195	10.4 ± 8.2	6.2 (12)	92.6 (90.3–94.5)	88.2 (84.2–92.2)
RTH	2	9.1 ± 5.5	0		100
RTH custom	2	9.5 ± 8.1	0		100
All RTH	4	9.3 ± 5.6	0		100
All conventional	283	11.0 ± 7.8	8.8 (25)	95.8 (94.4–97.2)	77.8 (73.1–82.5)
All custom	66	15.7 ± 7.1	9.1 (6)	91.7 (88.2–95.2)	89.7 (85.7–93.7)
All TKA	349	11.9 ± 7.9	8.9 (31)	95.0 (93.7–96.3)	81.6 (78.0–85.2)

CCK = constrained; CR = cruciate-retaining; PS = posterior-stabilized; RTH = rotating hinge.

which may be the result of an increased percentage of referrals of these difficult cases. Another limitation is the study's retrospective approach that depended on a questionnaire evaluation by telephone without clinical or radiologic evaluation. Nonetheless, this approach achieved followup of > 95%. There is also a degree of uncertainty in diagnosing juvenile idiopathic arthritis. There is a substantial number of alternative diagnoses that may be hard to differentiate from juvenile inflammatory arthritis other than by exclusion. In our study, diagnoses were determined from medical records and confirmed during telephone interviews. With many patients with juvenile inflammatory arthritis, TKA is hampered by the lack of appropriately designed implants and surgical instrumentation. The lack of components is not simply a result of patients with juvenile inflammatory arthritis being smaller. Simply scaling down adult TKA designs does not create the most viable solution. Differences in anatomy and function make designing juvenile inflammatory arthritis-specific medical devices challenging. Small sizes of commercial adult TKA systems often do not provide the alterations in shape required to fit the deformed anatomy of the juvenile inflammatory arthritis patient's knee and to restore adequate motion to the stiff joint.

Survivorship is likely impacted by the poor bone quality, joint deformities, and contractures with which these patients often presented and the immunosuppressive medications used to treat their disease. These procedures were performed by experienced surgeons who still consider these cases challenging. Patients with juvenile idiopathic arthritis have some of the most challenging deformities and bone abnormalities of any patient population. Few reports exist on TKA in patients exclusively with juvenile inflammatory arthritis, all with only small numbers of cases and mostly short-term followup (Table 1). Other studies report on young patients with rheumatoid arthritis with patients with juvenile inflammatory arthritis as a subset. In four of the studies on TKA for rheumatoid arthritis in patients under the age of 55 years, the number of knees ranged from only 25 to 103 and the length of followup ranged from 7 to 18 years [2, 5–7]. However, the revision rates were low with survivorship reported as high as 94% at 20 years [5]. Bellemans et al. [2] reported on 25 TKAs in patients younger than 45 years with rheumatoid arthritis followed for 7 years with excellent pain relief and functional improvement and a survival rate of 90% at 10 years. Dalury et al. reported 103 TKAs in patients younger than 45 years, 29% of whom had juvenile rheumatoid arthritis [6]. There were three revisions (3%) at 7.2 years followup. Crowder et al. [5] reported on 47 TKAs in 32 patients younger than 55 years (average age 43 years) with rheumatoid arthritis with an average 18-year followup. They reported six revisions for polyethylene wear but still estimated a survival rate of 100% at 15 years and 93.7% at 20 years. Gill et al. [7] reported on 68 TKAs (29 for rheumatoid arthritis) in patients younger than 55 years (average age 51 years). The revision rate was 3% at 9.9 years. Meding et al. [13] presented a consecutive series of 220 patients undergoing primary TKA for rheumatoid arthritis. Excluding infections (2.4%) and failed metal-backed patellar implants, survival rates were 99.5% at 10 years and 96.5% at 20 years. Instability was found in 15%. The studies that are limited to juvenile inflammatory arthritis have smaller numbers, shorter followup, higher revision rates, and lower survivorship when compared with the results of TKA for rheumatoid arthritis in the patients younger than 55 years (Table 1). These are small studies with the number of knees ranging from only 15 to 48,

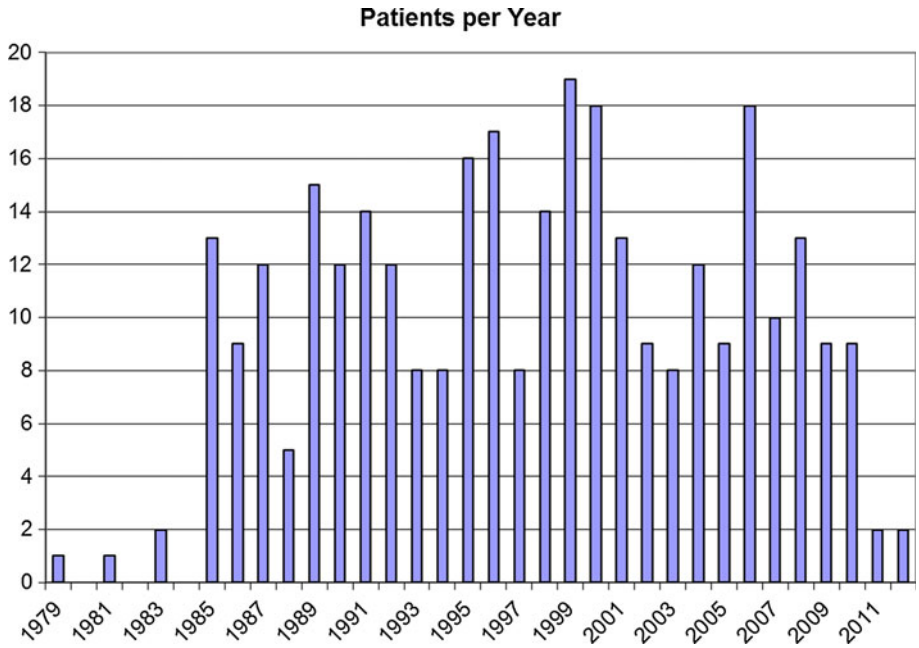

Fig. 5 Numbers of included TKAs in patients with juvenile idiopathic arthritis per year are shown.

whereas the length of followup ranged between 3.9 and 16 years. Whereas Carmichael and Chaplin [4] (25 knees) and Jolles and Bogoch [9] (22 knees) reported no revisions at early followup (5 and 8 years, respectively), the rest of the studies report revision rates from 4.5% at 3.9-year followup [3] to 20% at 15.5-year followup. Malviya et al. [12] presented 34 TKAs in 20 patients with survival rate of only 58% at 20 years. Our study has the largest number of knees (349) performed at one of the longest average followups (12.8 years) and illustrates the difference in survivorship of TKA in juvenile inflammatory arthritis as compared with the patients younger than 55 years with rheumatoid arthritis. There is little consensus in the literature about use of constraint in these patients, particularly over the long time period of this study. Accordingly, there was considerable variation on this issue among the participating centers (Table 3), and this may have contributed to differences in observed survivorship across the centers whose patients were included in this report (Table 2). The cruciate-retaining knees had superior survivorship (98.6%) as compared with the constrained (93.3%) and posterior-stabilized (91.7%) at 10 years, but surprisingly, the cruciate-retaining knees had the lowest survivorship at 20 years (69.2%). One would expect the more constrained knees to have higher failure rates for loosening and polyethylene wear, but the activity levels of the patients may have influenced the revision rates. The patients with greater deformities that might necessitate the use of greater constraint are more likely to be the less active patients resulting in lower stresses across the knees. In addition, in seven of the cruciate-retaining revisions, only the polyethylene insert was revised. Although this still was counted as an end point for knee survivorship, the original femoral and tibial components were not revised. The custom-fit knees were also typically used in the more severe cases with greater deformities and poorer functional abilities. The lower activity levels may explain why the results of the custom implants did not deteriorate significantly between 10 and 20 years.

The patients with juvenile inflammatory arthritis had significant functional limitations, which can be attributed to multiple joint involvement and preoperative functional deficiencies. Over 85% of the patients in this study had polyarticular disease, over 60% had previous hip arthroplasty, and 90% had bilateral knee involvement. As a result of their age, many patients with juvenile inflammatory arthritis postpone surgery, becoming nonambulatory or household ambulators by the time of surgery. Twelve percent of the patients were wheelchair-dependent at latest followup. Perhaps with improved medical treatments, patients will retain better functional status until they are older, and, if they do eventually undergo TKA, their disease may be more likely to be monoarticular, which would likely lead to better functional results. Surprisingly, the number of procedures performed each year remained relatively constant over the observation period despite the recent advent of new medications for treatment of juvenile inflammatory arthritis (Fig. 5). Although this may reflect an increase in the percentage of these cases referred to these institutions, changes in surgical indications over the 30-year period of the study may have impacted the yearly incidence of TKA in this patient cohort.

This study may help surgeons in counseling patients with juvenile inflammatory arthritis with realistic expectations toward outcome and survivorship of TKA. In patients with severe functional limitations despite conservative management, TKA may be indicated no matter how young the patient because extended periods of preoperative morbidity are associated with worse outcome and postoperative function [5, 10]. The surgeon needs to appreciate the challenging nature of these cases. Our current approach to the patient with juvenile inflammatory arthritis is to optimize their medical treatment preoperatively while also maximizing their physical therapy to maintain ROM. We no longer wait until the patient is nonambulatory to proceed with surgery. When patients have contractures in both knees, we prefer to perform simultaneous bilateral TKA to correct these deformities as long as the patients are fit for the larger bilateral procedure. We caution surgeons that these are technically difficult procedures; stiffness, flexion contracture, bony deformity, osteopenia, and small size all are common. The surgical exposure can be difficult in the severe deformities, because sometimes the patella is ankylosed to the femur and needs to be resected in situ. A lateral release early on may help with exposure in the contracted knee. More extensile exposures sometimes are called for, but tibial tubercle osteotomies often are not feasible if osteopenia is present or the tibia is small. To accurately determine if custom implants are needed, preoperative planning with CT of the knee can be very helpful. We attempt to maximize surface area contact to better distribute the stress in these patients with poor bone quality.

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