IN BRIEF

Primary Total Knee Arthroplasty in Hemophilia and Allied Disorders: Revision Rates and Their Causes

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Abstract

When patients with hemophilia and allied disorders (von Willebrand disease and other congenital bleeding disorders) do not receive adequate primary hematologic prophylaxis from infancy, their joints will suffer knee joint degeneration; when such joint degeneration becomes very advanced (painful and disabling) despite previous conservative treatment, the only way to alleviate the problem will be to implant a primary total knee arthroplasty (TKA). The literature has shown that twenty years after implantation, 71% of primary TKAs are still functional; on the other hand, 18% have to be revised as a consequence of periprosthetic joint infection (PJI). The main causes of revision total knee arthroplasty are PJI and aseptic loosening (39% each).

Level of evidence: III

Keywords: Hemophilia, Primary total knee arthroplasty, Revision rates, Von willebrand disease

Introduction

he aim of this article is to analyze what the literature published during the last seven years (2017-2023) says regarding survival rates of primary total knee arthroplasty (pTKA) and the frequency of revision TKA (rTKA) in patients with congenital bleeding disorders, hemophilia and von Willebrand disease (VWD), and their causes.

On 18 December 2023 a literature search was performed in PubMed using "hemophilia revision total knee arthroplasty" as keywords. Forty-one articles were found, of which 11 were finally analyzed (inclusion criteria) because they were all case series published between 2017 and 2023 presenting data on the parameters previously mentioned. The other 30 articles were excluded because they did not meet the aforementioned inclusion criteria.

Main body

In hemophilic patients pTKAs have a higher percentage of severe complications and different cumulative prosthetic survival rates than those implanted in patients with degenerative osteoarthritis or rheumatoid arthritis [Table 1]. Table 2 summarizes the case series published on rTKA

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in patients with congenital bleeding disorders [Table 2].¹⁻¹¹ Figure 1 shows the scattered plots of the average age of patients at the time of implantation of pTKA in papers published from 2017 to 2023 [Figure 1].

Table 1. Rates of severe complications and cumulative prosthetic survival after primary total knee arthroplasty (pTKA): comparison between hemophilic arthropathy, osteoarthritis and rheumatoid arthritis.

	SEVERE COMPLICATIONS	CUMULATIVE PROSTHETIC SURVIVAL
HEMOPHILIC ARTHROPATHY	22%	83%
OSTEOARTHRITIS	7%	98%
RHEUMATOID ARTHRITIS	9%	97%



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Table 2. Case se	ries published betwe	en 2017 and 2023 (on revision total k	mee arthroplasty (rT	'KA) in patients with h	nemophilia and allied disorders.
AUTHORS [REFERENCE]	YEAR / DISEASE	RATE OF rTKA	AGE (AVERAGE /RANGE) - YEARS	FOLLOW-UP (AVERAGE /RANGE) - YEARS	CAUSES of rTKA	PROSTHETIC SURVIVORSHI P OF PRIMARY TKA (pTKA)
Ernstbr unner et al [2]	2017 hemophilia	30% (13 / 43 pTKAs)	58 years (range, NA)	18 years (range, NA)	PJI (N=8); aseptic loosening (N=5)	The rate of survival free of revision (component removal for any reason) at 15 and 20 years for all 43 knees was 78% and 59%, respectively. Survivorship analysis with infection as the end point revealed a survival rate of 87% after 15 years and 82% after 20 years.
Song et al [3]	2017 hemophilia	2.3% (3 / 131 pTKAs)	41 years (range NA)	6.8 years (range NA)	PJI (N=3)	5-and 10-year survival rates were 98.5% and 97.7%, respectively
Santos Silva et al [4]	2019 hemophilia	0% (0 / 15)	39 years (range, NA)	11.3 years (range, NA)	rTKA was not required	Prosthesis survival rate was 94.4% at 11.3 years.
Bae et al [5]	2020 hemophilia	3.9% (3 / 102 pTKAs)	38.7 years (range, 26-69)	NA (mínimum 5 years)	PJI (N=2); aseptic tibial component loosening (N=1)	The Kaplan-Meier survivor graph showed 10- and 13-year prosthesis survival rates were 97.1% and 93.2%, respectively.
Kocaoglu et al [6]	2020 VWD	3.4% of pTKAs (NA)	64.9 years (range, NA)	5 years (range 12-10.8)	N=2, causes were not specified	NA
Li et al [1]	2020 hemophilia (N=78). This study compared OA, RA and hemophilia. Revisions rate: OA (2.8%), RA (7.14%), hemophilia (20.5%).	20.5% (16 / 78 pTKAs)	NA	6 years (range, NA)	N=16, causes were not specified	In this study, hemophilic arthropathy group had lower cumulative prosthetic survival rate (83%) when compared with OA or RA groups (98% or 97%). There was no significant difference between OA and RA groups.
Oyarzun et al [7]	2020 hemophilia	8.5% (5 / 60 pTKAs)	42 years (range, 24-75)	7 years (range, 5-11)	PJI (N=5)	92% at 5 years, 79% at 10 years
Goker et al [8]	2022 hemophilia. This study evaluated the effects of early major postop. bleeding on the implant survival of TKA in a single- center hemophilia cohort.	2.2% (1 / 45 pTKAs)	38 years (range, 18-63)	9.67 years (range, 2-22)	PJI (N=1). Eventually knee arthrodesis was required.	Kaplan-Meier analysis revealed a mean survival duration of 17 years for the bleeding group and 22 years for the control group. Survival rates were 80% for the bleeding group and 96% for the control group.
Carulli et al [9]	2023 hemophilia	2.2% (2 / 91 pTKAs)	39.3 years (range, 23-64)	12.3 years (range, 4.2-20.6)	Aseptic loosening (N=2)	The overall survivorship of the implants was 97.5% at the latest follow-up,

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REVISION RATES OF PTKA IN HEMOPHILIA

Table 2. Continu	ıed					
Feng et al [10]	2023 hemophilia	11.1% (4 / 36 pTKAs)	31.4 years (range, 18-54)	12.4 years (range, 10.2-17.3)	Aseptic loosening (N=4)	10- and 15-year prosthesis survival rates were 85.8% and 75.7%, respectively.
Liu et al [11]	2023 hemophilia	2.5% (2 / 71 pTKAs)	38.4 years (range, 21-63)	8.7 years (range, 4.3-15.2)	PJI (N=2)	With prosthetic revision as the endpoint, the 5- and 10-year survival rates of the prosthesis were 98.5% and 93.7%, respectively.

NA, not available; PJI, periprosthetic joint infection; OA, osteoarthritis; RA, rheumatoid arthritis; VWD, von Willebrand disease.

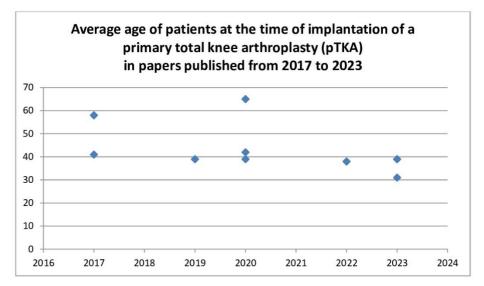


Figure 1. Scattered plots of average age (2017-2023)

Prosthetic survival rates of pTKA

Figure 2 shows the scattered plots of revision rates of pTKA in papers published from 2017 to 2023 [Figure 2].¹⁻¹¹ Survival rates can be measured by taking prosthetic revision as the end point or by taking infection as the end point. Both

figures clearly show how survival rates decrease over the years [Table 2, right column].¹⁻¹¹ Figure 3 shows the radiographic images of an aseptic loosening of a pTKA that required one-stage rTKA by means of a rotating hinge implant [Figure 3].

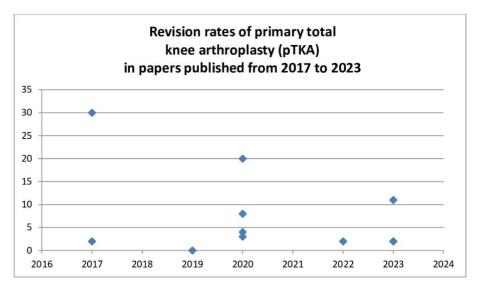


Figure 2. Scattered plots of revision rates (2017-2023)

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REVISION RATES OF PTKA IN HEMOPHILIA

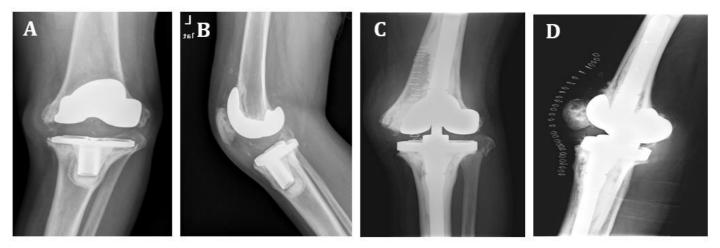


Figure 3 (A-D). Radiographic images of an aseptic loosening of a primary total knee arthroplasty (pTKA) that required one-stage revision total knee arthroplasty (rTKA) by means of a rotating hinge implant in a patient with hemophilia: (A) Preoperative anteroposterior (AP) radiograph; (B) Preoperative lateral view; (C) Postoperative AP radiograph; (D) Postoperative lateral view

Rates and Causes of rTKA

The rates of revision are shown in [Table 2, third column

from the left].

Figure 4 shows the causes of rTKA [Figure 4].¹⁻¹¹

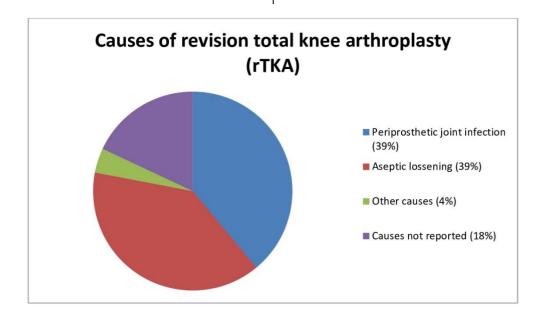


Figure 4. Causes of revision total knee arthroplasty (rTKA) in hemophilic patients

Conclusion

Twenty years after implantation, 71% of pTKAs are still functional in patients with hemophilia; however, 18% need to be revised as a result of PJI. The most frequent causes of rTKA are PJI (39%) and aseptic loosening (39%).

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REVISION RATES OF PTKA IN HEMOPHILIA

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