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Systematic Review

Outcomes of Lower Extremity Total Joint Arthroplasty in Patients With Skeletal Dysplasia: A Systematic Review

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ABSTRACT

Background: Patients with genetic skeletal dysplasias often require lower extremity total joint arthroplasty (TJA) due to early joint degeneration; however, little data exists regarding the outcomes of TJA in this population. Our purpose was to review the literature to determine the complication rates, revision rates, implant survivorship, and patient-reported outcomes of total knee arthroplasty and total hip arthroplasty (THA) in those with genetic skeletal dysplasias.

Methods: A systematic literature review of online databases (PubMed and Google Scholar) was conducted. Studies that reported the outcomes of THA or total knee arthroplasty in patients with genetically confirmed skeletal dysplasias were included. Case reports and studies that defined dysplasia based on height alone were excluded. Fourteen studies met the criteria for data extraction and analysis.

Results: Our review yielded a sample of 596 skeletal dysplasia patients with a median follow-up of 6.01 years (1.7-15.9). Mean age was 54.04 years, and mean body mass index was 29.1 kg/m^2. Cementless fixation was utilized in 65.7% of THAs, while all knees were cemented. Hip implant survivorship was 79% at 10 years and 56% at 20 years. Knee implant survivorship was 92% at 10 years and 46% at 20 years. Hip and knee revisions were 15.3% and 13.5%, respectively. The most common indication was aseptic loosening and polyethylene wear. Patient-reported outcomes improved across all domains.

Conclusions: The literature regarding lower extremity TJA in those with genetic skeletal dysplasias demonstrates appropriate 10-year implant survivorship and improvement in patient-reported outcomes across all survey domains.

Introduction and background

Skeletal dysplasia is a category of genetic conditions that cause atypical development of bones, joints, and cartilage. While a wide range of genetic alterations underlie these disorders, certain phenotypic traits such as short stature, hyperlordosis, and coxa vara are common in those with skeletal dysplasia [1-3]. In addition, skeletal dysplasia alters mechanical loading, weight distribution, bone quality, and joint configuration [4-7]. Collectively, these traits often trigger the development of early hip and/or knee degeneration, leading these patients to pursue early surgical intervention with orthopaedic providers [8,9]. While more conservative procedures such as femoral and acetabular osteotomies may offer temporary relief, skeletal dysplasia patients often require more definitive interventions including total knee arthroplasty (TKA) or total hip arthroplasty (THA) [10-13].

Lower extremity total joint arthroplasty (TJA) in this population requires fastidious preoperative planning to circumvent challenges posed by anatomic variances, small prostheses, poor bone quality, and severe joint deformities [2]. Despite efforts to optimize arthroplasty in this population, patients remain at substantial risk for complications such as intraoperative fracture, nerve injury, aseptic loosening, polyethylene wear, and joint stiffness [10,11,14-17]. As such, these patients often require subsequent revision surgeries, with small studies reporting revision rates of 4%-30% [4,10,11,16,17]. Some preliminary studies indicate that modular custom prostheses, cemented and cementless, may improve midterm implant survivorship [5,6,13,15,16,18,19]. However, the literature regarding long-term outcomes remains limited to small

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case series or studies that combine patients with short stature into a single analysis without a focus on skeletal dysplasia patients.

Due to the paucity of robust literature on this topic, a contemporary review and analysis of presently available data are indicated to provide additional guidance to those performing joint arthroplasty in this high-risk patient population. The purpose of this review was to determine the outcomes of TKA and THA in those with skeletal dysplasia including (1) complication rates; (2) incidence of revision arthroplasty; (3) implant survivorship; and (4) patient-reported outcomes (PROs). This is a contemporary analysis of TJA outcomes in skeletal dysplasia with a novel analysis of PROs based on the type of dysplasia.

Material and methods

Literature search and screening

A comprehensive literature review of the PubMed and Google Scholar online databases was conducted. The following keywords were used with AND or OR Boolean operators: ("Skeletal Dysplasia" OR "Achondroplasia" OR "Spondyloepiphyseal Dysplasia") AND (Arthroplasty OR Joint Replacement OR THA OR TKA). No additional query was utilized, and keywords were not employed in any alternative combination or permutation.

The initial query yielded 102 articles across both databases. After the exclusion of 8 duplicate results, studies were compiled for further screening by reviewers according to study criteria. The following inclusion criteria were applied: a full-text manuscript available in English pertaining to hip or knee arthroplasty in those with skeletal dysplasia confirmed by genetic testing; and a sample greater than 3 patients. Studies were excluded if they were not relevant to the topic, were case reports or background articles, defined dysplasia by height alone, or did not have full-text availability (Fig. 1).

Data acquisition

In the end, a total of 14 articles were included, and manuscripts were thoroughly evaluated for further data extraction (Table 1). Extracted data included study design, sample size, types of skeletal dysplasia, length of follow-up, surgical interventions, complication rate, revision rate, method of fixation, use of custom prostheses, implant survivorship, and PROs. Demographic data included mean age, height, weight, and body mass index (BMI). Survivorship was calculated from the subset of studies that reported survivorship outcomes from their cohort at the specified time point (<1 year, 5 years, 10 years). Patients were excluded from this analysis if the time points were not explicitly described.

Functional outcome measures

Due to discordance between studies regarding the choice of PRO surveys, results are reported both by survey type and by skeletal dysplasia type based on available data. Outcome measures utilized by investigators included Harris hip score (HHS), Oxford Hip Score, visual analog scale (VAS), Western Ontario and McMaster Universities Arthritis Index, Knee Society Score, and 12-item Short Form Survey. Additionally, one included study employed the Charnley, Merle d'Aubigné and Postel Method to evaluate THA patients postoperatively.

Data analysis

Descriptors were used to report individual study characteristics. Discrete variables were reported as counts or proportions. Normally distributed variables were reported as means with standard deviations or means with ranges based on reported data. Skewed variables were reported as medians with ranges.

Results

Study characteristics

In total, 596 skeletal dysplasia patients, who underwent 689 total joint replacements (367 THA and 322 TKA), were included across all 14 studies. All studies were retrospective in design. Twelve studies were case series, and 2 were cohort studies. Mean length of follow-up for studies ranged from 3.03 to 15.9 years. The most frequently reported types of dysplasia were achondroplasia (n = 442), spondyloepiphyseal dysplasia (SED) (n = 53), multiple epiphyseal dysplasia (MED) (n = 43), and osteogenesis imperfecta (OI) (n = 20) (Table 1).

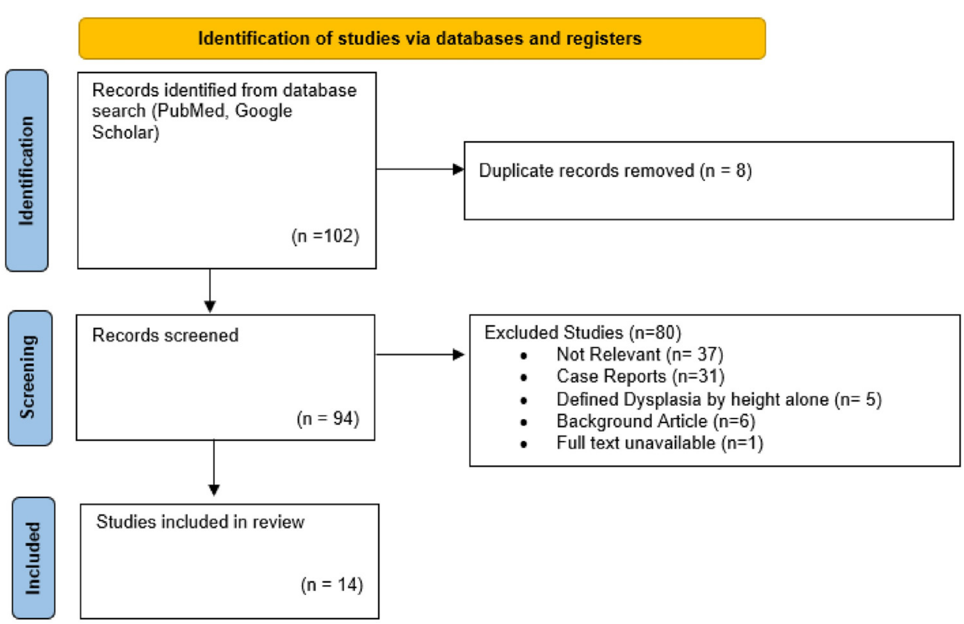

Figure 1. Study identification and screening process.

Table 1
Characteristics of included studies.

Authors	Year	Title	Level of evidence	Study design	Sample size	Type of dysplasia (n)
Moore et al. [20]	2021	Total Joint Arthroplasty in Patients With Achondroplasia: Comparison of 90-D Adverse Events and 5-Y Implant Survival	III	Retrospective-matched cohort	435	Achondroplasia (435)
Wyles et al. [21]	2019	Total Hip Arthroplasty Reduces Pain and Improves Function in Patients With Spondyloepiphyseal Dysplasia: A Long-Term Outcome Study of 50 Cases	IV	Retrospective case series	29	Spondyloepiphyseal dysplasia (29)
Osagie et al.[12]	2012	Custom total hip arthroplasty in skeletal dysplasia	IV	Retrospective case series	9	Diastrophic dysplasia (3), multiple epiphyseal dysplasia (2), Hurler syndrome (2), achondroplasia (1), pseudoachondroplasia (1)
Kim et al.[22]	2011	Technical challenges of total knee arthroplasty in skeletal dysplasia	IV	Retrospective case series	8	Achondroplasia (3), multiple hereditary exostosis (3), osteogenesis imperfecta (2)
Vanlommel et al. [23]	2017	Hybrid total hip arthroplasty for multiple epiphyseal dysplasia	IV	Retrospective case series	6	Multiple epiphyseal dysplasia (6)
Sewell et al. [24]	2012	Custom rotating-hinge primary total knee arthroplasty in patients with skeletal dysplasia	IV	Retrospective case series	8	Achondroplasia (2), spondyloepiphyseal dysplasia (1), pseudoachondroplasia (1), multiple epiphyseal dysplasia (1), Morquio syndrome (1), diastrophic sysplasia (1), Larsen's syndrome (1)
Sewell et al. [25]	2011	Custom cementless THA in patients with skeletal dysplasia results in lower apparent revision rates than other types of femoral fixation	IV	Retrospective case series	25	Multiple epiphyseal dysplasia (14), spondyloepiphyseal dysplasia (4), Morquio syndrome (2), diastrophic dysplasia (1), pseudoachondroplasia (1), osteogenesis imperfecta (1), Shwachman syndrome (1), Cornelia de Lange syndrome (1)
Ain et al. [5]	2004	Total hip arthroplasty in skeletal dysplasias: patient selection, preoperative planning, and operative techniques	IV	Retrospective case series	7	Spondyloepiphyseal dysplasia (3), diastrophic dysplasia (2), pseudoachondroplasia (1), Morquio syndrome (1),
Pavone et al. [26]	2009	Bilateral total hip arthroplasty in subjects with multiple epiphyseal dysplasia	IV	Retrospective case series	7	Multiple epiphyseal dysplasia (7)
Ke et al. [27]	2020	Short-term outcomes of total hip arthroplasty in the treatment of Tönnis grade 3 hip osteoarthritis in patients with spondyloepiphyseal dysplasia	IV	Retrospective case series	9	Spondyloepiphyseal dysplasia (9)
Raggio et al. [28]	2020	Joint replacements in individuals with skeletal dysplasias: one institution's experience and response to operative complications	III	Retrospective cohort	29	Spondyloepiphyseal dysplasia (7), pseudoachondroplasia (4), multiple epiphyseal dysplasia (4), diastrophic dysplasia (3), osteogenesis imperfecta (2), Kniest dysplasia (2), achondroplasia (1), Morquio syndrome (1), Hurler syndrome (1), Steel syndrome (1), spondyloepimetaphyseal dysplasia (1), metaphyseal dysplasia (1), unknown (1)
Ramaswamy et al. [29]	2010	Total hip replacement in patients with multiple epiphyseal dysplasia with a mean follow-up of 15 y and survival analysis	IV	Retrospective case series	9	Multiple epiphyseal dysplasia (9)
Krishnan et al. [30]	2013	Primary and revision total hip arthroplasty in osteogenesis imperfecta	IV	Retrospective case series	4	Osteogenesis imperfecta (4)
Carlson et al. [31]	2020	Total Hip Arthroplasty in Patients With Osteogenesis Imperfecta	IV	Retrospective case series	11	Osteogenesis imperfecta (11)

Patient demographics

Of 596 skeletal dysplasia patients, 227 were male (39.9%), and mean age was 54.04 years. BMI, height, and weight were reported for 146 patients with resulting means of 29.1 kg/m^2 (15.5-66.1), 141.1 cm (88.9-179), and 57.4 kg (24.9-113.4), respectively. Median length of follow-up for all patients was 6.01 years (1.7-15.9) (Table 2).

Implant design and survivorship

Method of fixation in 172 THAs was as follows: cementless (n = 113, 65.7%), cemented (n = 47, 27.3%), hybrid (n = 11, 6.4%), and reverse hybrid (n = 1, 0.6%). Hip prostheses utilized were custom (n = 89, 24.3%), not custom (n = 128, 34.9%), and unspecified (n = 150, 40.9%) (Table 2). Hip implant survivorship was 79% (44/56) at 10 years and 56% (28/50) at 20 years (Table 3).

All knees for which the TKA method of fixation was reported were cemented (n = 11, 100%). Knee implant design was as follows: custom (n = 25, 7.8%), not custom (n = 12, 3.8%), and unspecified (n = 285, 88.5%) (Table.2). Knee implant survivorship was 92% (12/13) at 10 years and 46% (6/13) at 20 years (Table 3).

Complications and revisions

THA complications and revisions were reported by 11 studies, which encompassed 217 procedures. The most common complications were heterotopic ossification (n = 22, 10.1%), polyethylene

Table 2
Demographics, method of fixation, and implant design of skeletal dysplasia patients.

Mean (range) age (y) (n = 596)	54.04 (14-80)
Sex (n = 596)	
Male (%)	227 (39.9%)
Female (%)	369 (61.9%)
Mean (range) weight (kg) (n = 117)	57.4 (24.9-113.4)
Mean (range) height (cm) (n = 146)	141.1 (88.9-179)
Mean (range) body mass index (kg/m^2) (n = 84)	29.1 (15.5-66.1)
Median (range) length of follow-up (y) (n = 596)	6.01 (1.7-15.9)
TKA method of fixation (n = 8; 11 knees)	
Cemented	11
THA method of fixation (n = 107; 172 hips)	
Cemented	47
Cementless	113
Other	12 (11 hybrid, 1 reverse hybrid)
Implant design	
Custom	114 (89 hips, 25 knees)
Not custom	140 (128 hips, 12 knees)
Not specified	435 (150 hips, 285 knees)

Table 3
Hip and knee implant survivorship and failure rate in skeletal dysplasia patients at 0, 10, and 20 years.

Outcomes	t = 0 y	t = 10 y	t = 20 y
Surviving hips/total hips reported at time point	367/367	44/56	28/50
Total hip survivorship %:	100%	79%	56%
Total hip failure %	0%	21%	44%
Surviving knees/total knees reported at time point	298/298	12/13	6/13
Total knee survivorship %:	100%	92%	46%
Total knee failure %:	0%	8%	54%

wear (n = 15, 6.9%), aseptic loosening (n = 12, 5.5%), acetabular osteolysis (n = 9, 4.1%), femoral osteolysis (n = 8, 3.7%), and intraoperative fracture (n = 8, 3.7%). Other complications included periprosthetic fracture (n = 5, 2.3%), deep infection (n = 5, 2.3%), and nerve injury (n = 3, 1.4%). Five nonrevision interventions occurred, including 3 removal of hardware and 2 irrigation & debridement (Table 4).

Of 367 primary THAs, 56 required revision surgery (15.3%). More specifically, revisions included liner exchange (n = 16, 4.4%), femoral component (n = 13, 3.5%), acetabular component (n = 8, 2.2%), both components (n = 6, 1.6%), and unspecified revision (n = 13, 3.5%). Reasons for revision were reported for 217 THAs. Top reasons for revision were aseptic loosening (n = 21, 9.7%), polyethylene wear (n = 15, 6.9%), and recurrent instability (n = 4, 1.8%) (Table 4).

TKA complication data were reported by 4 studies with a total sample of 322 joint replacements. The most frequent complications were the requirement of blood transfusion (n = 19, 5.6%), readmission (n = 16, 5.0%), urinary tract infection (n = 14, 4.3%), periprosthetic fracture (n = 2, 0.6%), and nerve injury (n = 2, 0.6%). Other notable complications were aseptic loosening (n = 1, 0.3%) and persistent knee pain (n = 1, 0.3%). No superficial or deep infections were reported. The only nonrevision intervention that was pursued was manipulation under anesthesia in 14 cases (4.3%) (Table 4).

Revision rates were provided by 3 studies spanning 37 TKAs. Five revisions were reported (13.5%) due to periprosthetic fracture (n = 2), aseptic loosening (n = 1), patellar tracking issues (n = 1), and persistent pain and stiffness (n = 1) (Table 4).

Patient-reported outcomes

When stratified by skeletal dysplasia type, PROs were available for MED, SED, and OI. MED patients who underwent THA had reduction in pain postoperatively, with mean VAS change from 7 to 1 (n = 6). In addition, Postel-Merle d'Aubigné score improved from a mean score of 7.63-14.23 (n = 15). SED patients who underwent THA had postoperative improvement in HHS from a mean of 44.78-87.5 (n = 38). Those with SED also had improvement in Western Ontario and McMaster Universities Arthritis Index pain, stiffness, and physical function scores (n = 9) (Table 5). Lastly, OI patients had improvement in Oxford Hip Score following THA with a mean score change from 15 to 41 (n = 4). OI group also improved in HHS after THA from a mean of 46-75 (n = 11) (Table 5).

For more inclusive analysis, all skeletal dysplasia types were combined based on the survey administered. HHS score improved from a mean of 43.76-82.12 following THA (n = 84, 129 THA). Pain based on VAS was reduced from a mean of 6.72-1.65 (n = 22, 31 THA). The 12-item Short Form Survey also improved from a mean of 41.56-56.33 (n = 22, 31 THA). For those undergoing TKA, Knee Society Score improved from a mean of 30.21-75.77. This trend was also consistent in the knee society function score, which improved from 34.56-74.37 postoperatively (n = 16, 23 TKA) (Table 5).

Discussion

Skeletal dysplasias are genetic disorders implicated in the development of aberrant bony and cartilaginous structures, resulting in a constellation of skeletal deformities. Phenotypic traits common to these conditions include short stature, hyperlordosis, and coxa vara, which result in atypical challenges during TJA [1,2]. Our systematic review of TJA in these challenging cases showed: (1) there was an increased utilization of cemented fixation or custom prostheses in this patient population relative to nonskeletal dysplasia populations due to their bone quality, short stature, and anatomical variation; (2) overall survivorship was lower than nonskeletal dysplasia populations; however, failure modes were relatively consistent with nonskeletal dysplasia populations, most commonly aseptic loosening/polyethylene wear and periprosthetic fracture; (3) improvements in commonly utilized patient-reported outcome measure scores suggest functional benefit of these surgeries in this patient population.

Given anatomic variances, poor bone quality, and severe limb or joint deformities, lower extremity TJA in this population is particularly challenging due to the increased risk of operative complications. Subjects in our study that underwent TJA had mean height, weight, and BMI of 141.1 cm, 57.4 kg, and 29.1 kg/m^2, respectively. In addition, patients underwent TJA at a mean age of 54 years, raising additional considerations for method fixation and survivorship. Among cases with reported method of fixation (172 THA, 11 TKA), cementless fixation was utilized in 66% of THAs (n = 113), whereas all TKAs were cemented. Custom prostheses were also utilized in 41% (n = 89) of hip arthroplasties and 67.6% (n = 25) of knee arthroplasties for which implant design was reported. For THA, implant sizes were not always reported, but femoral head sizes were frequently between 22 and 32 mm in diameter with acetabular component diameters between 40 and 52 mm when

Table 4
Complications and revisions of total hip and knee arthroplasty in skeletal dysplasia patients.

THA complications (n = 217 hips)	#	%	TKA complications (n = 322 knees)	#	%
Heterotopic ossification	22	10.1%	Heterotopic ossification	0	
Polyethylene wear	15	6.9%	Aseptic loosening	1	0.3%
Aseptic loosening	12	5.5%	Osteolysis	0	-
Acetabular osteolysis	9	4.1%	Intraoperative fracture	0	-
Femoral osteolysis	8	3.7%	Periprosthetic fracture	2	0.6%
Intraoperative fracture	8	3.7%	Tibial	1	0.3%
Femur	6	2.8%	Patellar	1	0.3%
Acetabulum	2	0.9%	Deep infection	0	-
Periprosthetic fracture	5	2.3%	Superficial infection	0	-
Deep infection	5	2.3%	Nerve injury/palsy	2	0.6%
Superficial infection	2	0.9%	Pulmonary embolism	0	-
Nerve injury/palsy	3	1.4%	Pneumonia	0	-
Pulmonary embolism	3	1.4%	Migration	0	-
Pneumonia	3	1.4%	Deep vein thrombosis	0	-
Migration	2	0.9%	Persistent pain	1	0.3%
Deep vein thrombosis	2	0.9%	Transfusion	18	5.6%
Persistent pain	2	0.9%	Dislocation	0	-
Transfusion	1	0.5%	Hemorrhage/hematoma	0	-
Dislocation	1	0.5%	Dehiscence	0	-
Hemorrhage/hematoma	1	0.5%	Urinary tract infection	14	4.3%
Dehiscence	0	-	Readmission	16	5.0%
Urinary tract infection	0	-	Nonrevision intervention	14	4.3%
Nonrevision intervention	5	2.3%	MUA	14	4.3%
Removal of hardware	3	1.4%	Removal of hardware	0	-
I&D	2	0.9%	I&D	0	-
THA revisions (n = 367 hips)	**#**	**%**	**TKA revisions (n = 37 knees)**	**#**	**%**
Total revisions	56	15.3%	Total revisions	5	13.5%
Liner exchange	16	4.4%	Reasons for revision		
Femoral	13	3.5%	Periprosthetic fracture	2	5.4%
Acetabular	8	2.2%	Aseptic loosening	1	2.7%
Both components	6	1.6%	Patellar tracking issues	1	2.7%
Unspecified	13	3.5%	Persistent pain/stiffness	1	2.7%
Reasons for revision (n = 217 hips)					
Aseptic loosening	21	9.7%			
Polyethylene wear	15	6.9%			
Recurrent instability	4	1.8%			
Migration	3	1.4%			
Periprosthetic fracture	3	1.4%			
Periprosthetic infection	1	0.5%			
Persistent pain	1	0.5%			
Other/unspecified	8	3.7%			

MUA, manipulation under anesthesia; I&D, irrigation and debridement.

reported [2,15,16,18]. Cumulative rate of revision for hip and knee arthroplasty was 15% and 13.5%, respectively. 10-year hip and knee survivorship were 79% and 92%, respectively, while 20-year hip and knee survivorship were 56% and 46%, respectively.

Optimizing survivorship has been explored by a small preliminary case series, some of which displayed promising results with cementless modular custom prostheses [5,6,13,16,19,22]. While early studies emphasized cement fixation for custom components to accommodate for variations in canal size and poor bone quality [5], later studies utilizing press-fit prostheses had favorable results [2,16]. A study by Lim et al. of 23 THAs in those with MED using modular cementless prostheses revealed no hips required revision due to aseptic loosening of either component. Only one hip required revision due to polyethylene wear and osteolysis (4.3%). They reported only 3 complications including heterotopic ossification (8.7%) and only one intraoperative femoral fracture (4.3%), which occurred at the time of press-fit stem insertion [16]. Lim et al. suggested that the modular component offered enhanced the intraoperative maneuverability and accuracy of femoral offset [2,16], leading to a reduction in periprosthetic fracture without a resultant increase in loosening. This study, while small, supports utilization of cementless modular components to reduce rates of aseptic loosening, which was a leading reason for revision in our study. These results were further supported by Osagie et al., a series of 14 THAs in those with genetic dysplasias, which utilized press-fit custom prostheses in 12 hips without any intraoperative fractures. Notably, 2 of the 3 cemented components required revision for aseptic loosening, leading the authors to suggest that the stability offered by press-fit components was vital in this population due to variations in pelvic loading [2]. In contrast to a skeletal dysplasia population, the cumulative revision rates for osteoarthritis patients undergoing primary THA and TKA are approximately 3% after 5 years and 4% after 10 years, based on data from the Australian Orthopaedic Association National Joint Replacement Registry [32]. This suggests that the surgical challenges and comorbidities of patients with skeletal dysplasia appear to adversely affect survivorship relative to other primary osteoarthritis populations.

Early series also noted high rates of periprosthetic femoral fractures, which were attributed to severe femoral deformities [5,6,10,11]. In addition, preliminary studies also revealed a concerning rate of nerve injury, which contemporary investigators have associated with aggressive soft tissue release and difficulty gaining exposure [2,22]. In contrast, our study revealed the most common complications following THA were heterotopic ossification (10.1%, n = 22), polyethylene wear (6.9%, n = 15), and aseptic loosening (5.5%, n = 12), the rates of which were more consistent with more recent analyses [2,16]. Only eight cases (3.7%) were complicated by intraoperative fractures (6 femoral and 2 acetabular), and 3 patients (1.4%) suffered nerve injuries. Five patients had periprosthetic fractures postoperatively (2.3%). The most common complications associated with TKA were the requirement for blood transfusion (5.6%, n = 18) and urinary tract infection (4.3%, n = 14). The use of highly cross-linked polyethylene was not always explicitly described and may have accounted for higher rates of polyethylene wear in older studies.

Despite inconsistencies regarding method of fixation, implant design, and operative approach, prior series and the results of the present study are concordant regarding the positive effect of TJA based on PROs. Patients showed vast improvement in scores across all surveys, which remained consistent when further stratified by type of dysplasia. The benefits of lower extremity TJA in those with genetic skeletal dysplasias are undeniable, given the marked improvement across all survey domains including function, pain, and stiffness.

Our study was primarily limited due to a lack of prior literature regarding lower extremity TJA in those with genetic skeletal dysplasias. Included studies were primarily retrospective in design with small sample sizes and inconsistent reporting across studies, which precludes meta-analysis. In addition, length of follow-up for many studies was inadequate for 10- and 20-year survivorship analysis, and individual patients' time of failure was not always explicitly described, limiting the survivorship analysis. Complication rates may also be underrepresented, as investigators differed in their choice of reported measures. PROs were similarly limited due to a lack of standardization in the employed surveys. Furthermore, skeletal dysplasias were largely grouped for analysis, which may obscure key differences in underlying pathologies.

Table 5
Patient-reported outcomes after lower extremity total joint arthroplasty by type of survey and type of skeletal dysplasia.

Survey name	Number of patients and joints	Mean preoperative score (range or SD)	Mean postoperative score (range or SD)	Mean difference	Percent change
HHS	83 patients; 129 THA	43.76 (24-72)	82.12 (39-100)	38.36	87.66
OHS	4 patients; 6 THA	15 (14-24)	41 (37-46)	26	173.33
MdAP	15 patients; 26 THA	7.63 (2-11)	14.23 (5-18)	6.6	86.50
VAS	22 patients; 31 THA	6.72 (SD = 1.59)	1.65 (SD = 1.55)	−5.07	−75.45
SF-12	9 patients; 12 THA	41.56 (SD = 4.22)	56.33 (SD = 3.3)	14.77	35.54
WOMAC	16 patients; 21 THA	-	-	-	-
Pain		12.94 (SD = 2.5)	5.63 (SD = 3.6)	−7.31	−56.49
Stiffness		4.7 (SD = 1.47)	2.54 (SD = 1.07)	−2.16	−45.96
Function		44.73 (SD = 8.93)	27 (SD = 9.33)	−17.73	−39.64
Knee Society	16 patients; 23 TKA	-	-	-	-
KSS		30.21 (14-60)	75.77 (28-90)	45.56	150.81
Function		34.56 (5-60)	74.37 (22-100)	39.81	115.19

Dysplasia type	Survey name	Mean preoperative score (range or SD)	Mean postoperative score (range or SD)	Mean difference	Percent change
MED (n = 6)	VAS	7 (not reported)	1 (not reported)	−6	−85.71
MED (n = 15)	MdAP	7.63 (2-11)	14.23 (5-18)	6.6	86.50
SED (n = 38)	HHS	44.78 (24-72)	87.5 (51-94)	42.72	95.40
SED (n = 9)	WOMAC				
	Pain	12.22 (SD = 1.9)	7.44 (SD = 2.96)	−4.78	−39.12
	Stiffness	4.56 (SD = 1.13)	3.11 (SD = 0.93)	−1.45	−31.80
	Physical Function	39.78 (SD = 5.52)	30.56 (SD = 5.32)	−9.22	−23.18
OI (n = 4)	OHS	15 (14-24)	41 (37-46)	26	173.33
OI (n = 11)	HHS	46 (35-59)	75 (47-97)	29	63.04

HHS, Harris hip score; OHS, Oxford Hip Score; MdAP, Merle d'Aubigné and Postel Score; VAS, visual analog scale; SF-12, 12-item Short Form Survey; WOMAC, Western Ontario and McMaster Universities Arthritis Index; KSS, Knee Society Score; MED, multiple epiphyseal dysplasia; SED, spondyloepiphyseal dysplasia; OI, osteogenesis imperfecta.

Conclusions

A review of literature regarding lower extremity primary TJA in those with genetic skeletal dysplasias revealed marked improvement in PROs across all survey domains, appropriate 10-year implant survivorship, and hip and knee revision rates of 15.3% and 13.5%, respectively, most commonly due to aseptic loosening and polyethylene wear.

Conflicts of interest

B. F. Ricciardi received research funding from Johnson and Johnson, is an editorial board member of Clinical Orthopaedics and Related Research, HSS Journal, The Knee, Arthroplasty Today, and is a board/committee member of AAOS-Hip Program Committee; AAOS-Practice Management Program Committee; AAHKS-Quality Measures Committee. All other authors declare no potential conflicts of interest.

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CRediT authorship contribution statement

Paul Guirguis: Writing – review & editing, Writing – original draft, Visualization, Methodology, Data curation, Conceptualization. **Lucas Fowler:** Writing – review & editing, Writing – original draft, Visualization, Methodology, Data curation, Conceptualization. **Benjamin F. Ricciardi:** Writing – review & editing, Writing – original draft, Visualization, Supervision, Methodology, Data curation, Conceptualization.

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