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Total Hip Arthroplasty in Sickle Cell Disease: Implications and Challenges

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Abstract

Sickle cell disease (SCD) is one of the most common inherited hemoglobinopathies in the West African sub-region and is frequently complicated by musculoskeletal manifestations, especially avascular necrosis (AVN) of the femoral head. This complication can progress to femoral head collapse and secondary osteoarthritis, leaving total hip arthroplasty (THA) as the only definitive solution. However, THA in patients with SCD is fraught with unique challenges: altered bone morphology, poor bone quality, increased risk of intraoperative fracture, susceptibility to infection, and a higher likelihood of revision compared to standard populations. This review synthesizes the available literature on the clinical indications, perioperative optimization, surgical considerations, implant selection, and outcomes of THA in SCD. It highlights both the advances achieved and the persistent barriers to optimal outcomes, particularly in resource-limited regions where the disease burden is greatest. While complications remain more frequent than in the general population, THA in SCD continues to demonstrate significant pain relief, improved function, and enhanced quality of life when performed in specialized settings with multidisciplinary care.

Subject Areas

Orthopedics

Keywords

Sickle Cell Disease, Total Hip Arthroplasty, Avascular Necrosis

1. Introduction

Sickle cell disease (SCD) is a disorder resulting from the inheritance of abnormal hemoglobin S. It is a condition with significant global health impact. When oxygen levels decrease, hemoglobin S polymerizes, forming chains that deform red blood cells into a sickle shape. These sickle-shaped cells are fragile, leading to hemolysis, and they can obstruct small vessels, contributing to both hemolytic and vaso-occlusive complications [1]. This pathophysiology underlies the myriad problems associated with SCD, including painful crises, organ damage, and bone pain from ischemia [1]. Worldwide, more than three hundred thousand infants are born annually with SCD. Advances in newborn screening and supportive care have improved survival, allowing many patients to live into adulthood [2] [3]. Consequently, long-term complications have gained greater clinical importance, with orthopedic sequelae being particularly significant [4] [5].

Among the musculoskeletal complications, avascular necrosis (AVN) of the femoral head is notably the most disabling. Recurrent vaso-occlusion compromises blood flow to the femoral head, leading to subchondral bone death, structural collapse, and subsequent degenerative damage to the hip joint [6]. Studies report a lifetime risk of AVN that can reach 50% for patients with hemoglobin SS disease, with many cases manifesting in the second to fourth decades of life. Notably, more than half of hip AVN cases are bilateral [7] [8]. Nonoperative strategies, including bisphosphonates, core decompression, or bone grafting, may provide benefit in early stages. However, these modalities generally offer only short-term relief and become ineffective once femoral head collapse occurs [9] [10]. For advanced AVN, total hip arthroplasty (THA) remains the definitive treatment, reliably offering pain relief and functional restoration [11].

The decision to perform THA in SCD is complex. Patients are often younger than the typical osteoarthritis population, raising concerns about implant longevity. Technical challenges, such as altered femoral canal anatomy and poor bone stock, combined with systemic risks like infection and vaso-occlusive crises, make THA in SCD particularly demanding [11]-[13]. This narrative review examines the outcomes and challenges of THA in SCD, with a focus on perioperative care, surgical techniques, clinical results, and future directions.

2. Epidemiology of AVN in Sickle Cell Disease

Avascular necrosis (AVN) of the femoral head is a common musculoskeletal complication of SCD. Overall, AVN occurs in approximately ten percent of SCD cases, but the risk for patients with HbSS genotype can rise to about fifty percent by mid-adulthood. The condition frequently presents bilaterally and typically manifests at a young age, usually within the second to fourth decade of life [7] [14]. The primary etiology is ischemic, stemming from repeated vaso-occlusive episodes that damage the vascular supply to the femoral head [1] [6] [15]. Additional risk factors include high baseline hemoglobin S levels, a history of acute chest syndrome, and corticosteroid use [16] [17].

The prevalence of AVN varies geographically. Studies from sub-Saharan Africa report that 20 to 50 percent of SCD patients have AVN [18] [19]. Improved survival into adulthood, attributable to newborn screening and hydroxyurea therapy, has increased the clinical relevance of AVN, as patients now live long enough to develop end-stage hip disease [3] [4] [20].

3. Indications for Total Hip Arthroplasty in SCD

The primary indication for THA is symptomatic AVN with femoral head collapse or secondary arthritis [11] [21]. Nonoperative strategies, including bisphosphonates, core decompression, and bone grafting, may offer temporary relief in early-stage disease but generally fail to prevent progression [9] [10] [22].

Patient Profile Differences

SCD patients undergoing THA differ from typical osteoarthritis populations:

- **Younger age:** Mean age 20 - 40 years versus 65 - 75 years in degenerative OA [11] [23].
- **Bilateral involvement:** Up to 40% require bilateral THA during their lifetime, sometimes simultaneously [24].
- **High functional demands:** Younger patients often seek rapid return to work and active lifestyles, emphasizing the need for durable implants.

Table 1 summarizes the demographic profile and outcomes of SCD patients undergoing THA compared with OA patients.

Table 1. Comparative demographics and outcomes of SCD vs OA THA patients.

Parameter	SCD Patients	OA Patients	Evidence/Sources
Mean age at THA	~20 - 35 years (often 30 - 35 yrs) (PubMed)	~65 - 75 years (typical primary OA cohort)	Typical in OA literature (textbook standard) [11]
Bilateral THA (%)	~21% - 40% (multiple cohorts show high rates of bilateral hip disease) (PubMed) [25]	~5% - 10%	Generally low in OA because disease tends to be unilateral or asymmetrical
Average Pre-op HHS	~20 - 45 (varies by series) (Lippincott Journals) [26]	~55 - 65 at presentation	Based on OA functional outcomes in multiple joint registries
Periprosthetic Joint Infection (2-year)	~5.0% (higher than OA) (PubMed) [27]	~2.5% (typical)	From national database comparison
2-year Dislocation Rate	~1.7% - 3.7% (higher than OA) (index.mirasmart.com) [28]	~0.8%	OA registries report low early dislocation rates
2-year Aseptic Loosening	~1.9% (higher than ON non-SCD) (PubMed) [27]	~0.68% (in non-SCD ON or OA)	Pooled database analyses

4. Perioperative Challenges

THA in SCD presents unique perioperative challenges due to hematologic, anesthetic, and orthopedic factors. Table 2 shows the common perioperative complications in SCD THA.

4.1. Hematological Considerations

Perioperative management aims to minimize sickling crises triggered by hypoxia,

Table 2. Common perioperative complications in SCD THA.

Complication	Incidence (%)	Notes
Vaso-occlusive crisis	9 - 19	Triggered by hypoxia, dehydration [29]
Acute chest syndrome	1.3 - 14	Reduced with pre-op transfusion [29]
Intraoperative femoral fracture	5 - 10	Higher in cementless THA [12]
Periprosthetic joint infection	6 - 10	Immunocompromised, functional asplenia [27]
Dislocation	3 - 7	Slightly higher than general population [28]

acidosis, hypothermia, or dehydration [30]. Preoperative exchange transfusions to reduce HbS levels below 30% reduce vaso-occlusive episodes and acute chest syndrome [21] [31] [32], while careful balancing minimizes alloimmunization and iron overload risks [33]. SCD's hypercoagulable state also necessitates thoughtful thromboembolism prophylaxis [32].

4.2. Anesthetic Management

Both regional and general anesthesia have advantages and risks. Maintaining normothermia, hydration, adequate oxygenation, and avoidance of acidosis is universally critical [1] [2] [34] [35].

4.3. Bone Quality and Anatomy

Sclerotic femoral canals, acetabular cysts, and osteopenia complicate fixation and increase risks of fracture and loosening [36]-[38].

4.4. Infection Risk

SCD patients face approximately double the risk of periprosthetic joint infection compared to controls, attributed to functional asplenia and chronic anemia [27] [28].

5. Surgical Strategies for THA in SCD

5.1. Fixation Methods

Cemented, uncemented, and hybrid fixation strategies each have roles, with increasing preference for cementless implants due to superior long-term survivorship [39] [40].

5.2. Bearing Surfaces

Ceramic-on-ceramic and highly cross-linked polyethylene bearings are favored for younger patients due to reduced wear and osteolysis [40]-[42].

5.3. Surgical Approach

Evidence suggests the anterolateral approach yields lower complication and dislocation rates in SCD patients [29].

6. Outcomes of THA in SCD

6.1. Functional Improvement

Harris Hip Scores typically improve from <40 preoperatively to >85 postoperatively, with substantial gains in pain relief and quality of life [11] [26] [43] [44].

6.2. Implant Survival

Modern cementless techniques report survivorship exceeding 90% at 10 - 15 years in selected cohorts, though revision risk remains higher than in OA populations.

7. Discussion

Total hip arthroplasty (THA) in patients with sickle cell disease (SCD) represents the intersection of a serious systemic illness and a complex surgical procedure. This review has synthesized studies covering the indications, perioperative considerations, surgical strategies, and outcomes of THA in this population. A discussion is necessary to contextualize these findings, clarify discrepancies in the literature, and provide guidance for clinicians, especially those working in high-burden, resource-limited settings.

7.1. SCD-Specific Pathophysiology and Its Surgical Implications

The fundamental disease process of SCD, involving repeated hemoglobin S polymerization, erythrocyte sickling, vaso-occlusion, and chronic hemolysis, directly and indirectly affects THA outcomes. Recurrent ischemia causes not only AVN of the femoral head but also widespread alterations in bone quality, including sclerosis, cyst formation, marrow hyperplasia, and osteopenia [1] [6] [36] [38]. These changes distinguish SCD-related hip disease from osteonecrosis from other etiologies and primary osteoarthritis, and help explain the higher rates of intraoperative complications and late failures observed in this population.

The femoral canal in SCD patients is often narrow, irregular, and sclerotic due to persistent marrow hyperplasia and infarcts. This morphology increases the risk of canal perforation, femoral fracture, and component malalignment during broaching, particularly when using cemented stems [36]. Similarly, the acetabular bone often exhibits cysts and segmental defects secondary to collapse and wear, complicating cup fixation and raising the risk of early loosening if adequate fixation is not achieved [37]. This altered anatomy underscores the necessity for meticulous preoperative planning, including templating, having a wide range of implant sizes available, and being prepared to alter fixation strategies intraoperatively.

Beyond bony anatomy, the systemic effects of SCD—functional asplenia, chronic anemia, endothelial dysfunction, and a prothrombotic state—significantly impact perioperative risk. Functional asplenia increases susceptibility to infection, while chronic anemia limits physiological reserve. Endothelial activation and hypercoagulability elevate the risk of thromboembolism, even in young patients [32]. Collectively, these factors contribute to higher rates of periprosthetic joint infection, medical complications, and revision surgery compared with non-SCD populations [27]-[29].

7.2. Indications for THA and Timing of Intervention

The primary indication for THA in SCD is typically AVN with femoral head collapse and secondary osteoarthritis, a pattern consistent across geographic regions. The optimal timing of THA in SCD remains debated. Patients often develop symptomatic AVN in their early twenties or thirties, long before the typical age for degenerative hip disease, raising concerns about implant longevity, the inevitability of revision surgery, and the cumulative health burden of multiple procedures over a lifetime.

Despite these concerns, delaying THA until late-stage disease can be counterproductive. Protracted pain, immobility, and deformity can lead to muscle atrophy, bone loss, and technically more difficult surgery. Several studies indicate that patients undergoing THA before the development of severe deformity and extensive acetabular bone loss achieve better functional outcomes and fewer complications [11] [26] [43]. Therefore, decision-making should not rely on age alone but should incorporate symptom severity, functional impairment, radiographic stage, and the patient's overall medical optimization.

7.3. Perioperative Optimization: Cornerstone of Success

Perioperative management is a critical determinant of outcome for THA in SCD. The literature indicates that many serious complications—such as vaso-occlusive crises, acute chest syndrome, and infection—are precipitated or exacerbated by physiological stressors including hypoxia, dehydration, acidosis, and hypothermia [30] [34] [35] [29].

Preoperative transfusion strategies have evolved. Early practices employed aggressive exchange transfusions to reduce hemoglobin S levels below 30%, which lowered the risk of acute chest syndrome and vaso-occlusive crises but increased risks of alloimmunization and iron overload [21] [31] [33]. Recent evidence supports a more tailored approach, balancing risks and often aiming for a modest increase in total hemoglobin rather than adhering to a strict hemoglobin S threshold, particularly in stable patients [30] [33].

Anesthetic management is complex. Both general and regional anesthesia can be used safely when managed by an experienced team adhering to physiological goals. Regional anesthesia may offer advantages in reducing blood loss and postoperative pain. Care must be taken to avoid hypotension to prevent tissue hypoxia and subsequent sickling [34]. Regardless of anesthetic technique, maintaining normothermia, hydration, oxygenation, and normal acid-base balance are essential tenets of care [1] [35].

Multidisciplinary collaboration among surgeons, hematologists, anesthesiologists, and infectious disease specialists is a key factor in improving outcomes [29] [45]. This approach is particularly important in high-burden regions, where delays in diagnosis, limited access to subspecialty care, and constrained resources may otherwise compound risk.

7.4. Implant Fixation: Cemented versus Cementless Debate

The choice between cemented and cementless fixation remains one of the most debated aspects of THA in SCD. Historically, cemented stems were favored due to concerns about achieving stable press-fit fixation in sclerotic femoral canals. Early series reported acceptable short-term outcomes; however, longer-term follow-up revealed relatively high rates of aseptic loosening, likely related to poor cement–bone interdigitation in abnormal bone [39].

Contemporary literature increasingly supports the use of cementless fixation, particularly with modern stem designs and refined surgical technique. Several series report excellent mid- to long-term survivorship exceeding 90% at 10 - 15 years with uncemented implants [39] [40]. These outcomes are attributed to biological fixation and avoidance of the compromised cement–bone interface seen in SCD. Nevertheless, the risk of intraoperative fracture is higher with cementless stems, emphasizing the need for gentle broaching, prophylactic cerclage wiring in high-risk cases, and readiness to alter the surgical plan if necessary [12] [36].

Hybrid fixation—cemented stem with an uncemented cup—has been proposed as a compromise in cases of severely compromised femoral bone. While outcomes are variable, this approach may be appropriate in selected patients, highlighting the importance of individualized decision-making rather than rigid adherence to a single fixation philosophy [46].

7.5. Bearing Surfaces and Dislocation Risk

Given the young age and high activity levels of many SCD patients undergoing THA, bearing surface selection is critical. The goal is to minimize wear, reduce osteolysis, and prolong implant longevity. Ceramic-on-ceramic and metal-on–highly cross-linked polyethylene bearings have emerged as preferred options, offering low wear rates and favorable long-term performance [41] [42]. Metal-on-metal bearings, once attractive for young patients, have been largely abandoned due to concerns regarding systemic metal ion toxicity and local adverse tissue reactions [41].

Dislocation rates in SCD patients are modestly higher than in primary osteoarthritis cohorts, reflecting a combination of altered anatomy, muscle weakness, and higher functional demands [28] [29]. Recent evidence suggests that the anterolateral surgical approach is associated with lower dislocation and overall complication rates compared with the posterior approach in this population [29]. Additionally, the increasing use of dual-mobility constructs may further mitigate instability risk, particularly in high-risk patients, although long-term data specific to SCD remain limited [47].

7.6. Functional Outcomes and Quality of Life

Despite the elevated risk profile, functional outcomes following THA in SCD are consistently favorable. Multiple studies demonstrate dramatic improvements in Harris Hip Scores, often rising from severely impaired preoperative levels (<40) to excellent postoperative scores (>85) within the first year [11] [26] [43]. These gains translate into meaningful improvements in mobility, independence, and overall quality of life, outcomes that are particularly impactful for young adults who may otherwise face decades of disability.

Importantly, patient-reported outcomes often emphasize pain relief and restoration of basic function rather than return to high-impact activities. This distinction underscores the importance of realistic preoperative counseling, ensuring that patients understand both the transformative potential of THA and the ongoing need for implant protection and follow-up.

7.7. Complications, Revisions, and Long-Term Survivorship

Although functional gains are substantial, complication and revision rates remain higher in SCD patients than in the general THA population. Infection, aseptic loosening, periprosthetic fracture, and medical complications account for most failures [27]-[29]. Aseptic loosening remains the dominant mode of late failure, reflecting both altered bone biology and the long life expectancy of these patients.

These realities necessitate long-term surveillance and a willingness to intervene early when problems arise. They also highlight the importance of implant choice and surgical technique aimed at maximizing initial stability and biological fixation.

7.8. Global Disparities and Future Directions

The burden of SCD is greatest in sub-Saharan Africa, where access to advanced arthroplasty care is often limited [18]. Paradoxically, much of the high-quality outcome data originates from high-income countries with lower disease prevalence. Addressing this disparity requires investment in regional centers of excellence, training in complex arthroplasty techniques, and access to affordable, durable implants suited to young patients.

Future directions are promising. Disease-modifying therapies such as voxelotor and crizanlizumab may reduce vaso-occlusive crises and potentially slow the progression of AVN [48]. Advances in biologic treatments and regenerative strategies may delay or obviate the need for arthroplasty in selected patients [49]. On the surgical front, computer-assisted navigation, robotics, and dual-mobility designs offer potential improvements in precision and stability, although their cost-effectiveness in low-resource settings must be carefully considered [50] [47].

7.9. Synthesis and Clinical Implications

In synthesis, THA in SCD is neither contraindicated nor futile; rather, it is a high-stakes intervention that demands meticulous planning, technical expertise, and multidisciplinary collaboration. When these elements are in place, outcomes can approach those seen in other complex arthroplasty indications, delivering profound and durable benefits to patients who would otherwise face severe lifelong disability.

The expanded body of evidence supports a nuanced approach: timely intervention, individualized perioperative optimization, preference for cementless fixation with modern bearings, and vigilant long-term follow-up. Continued research, particularly from high-burden regions, is essential to refine best practices and ensure equitable access to this life-changing procedure.

8. Future Directions

Advances in disease-modifying therapies, biologics, robotic-assisted surgery, and dual-mobility constructs may further improve outcomes. Addressing global disparities remains essential through multidisciplinary care and regional centers of excellence [45] [47]-[50].

9. Conclusion

Total hip arthroplasty in SCD is a high-risk yet transformative intervention. Despite higher complication rates than in osteoarthritis populations, THA reliably delivers profound functional improvement and pain relief when performed with meticulous planning, appropriate implant selection, and multidisciplinary perioperative care.

Conflicts of Interest

The authors declare no conflicts of interest.

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