

Cementless Total Hip Arthroplasties in Gaucher Disease: Long-Term Follow-Up

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ABSTRACT

We retrospectively assessed the clinical and radiologic outcomes of 15 primary cementless total hip arthroplasties performed in 12 young adults with type I Gaucher disease.

Mean Harris Hip Score was 86.9 points; patient satisfaction was high. There were no serious post-operative complications. Weight-bearing ambulation was immediate. Only 3 hips required revisions.

In cases of Gaucher disease, cementless hip arthroplasties are safe and effective. Because patients with chronic metabolic disorders differ from healthy patients with traumatic fractures, the results reported in this article may have implications for nontraumatic hip arthroplasty.

Gaucher disease, the most common lysosomal storage disease, is characterized by the presence of Gaucher cells in the spleen, liver, and bone marrow.¹ Since the advent of enzyme replacement therapy as treatment for this disease,² patients' quality of life has improved substantially, but the response of the bones lags behind that of the viscera.³

In 1966, Amstutz and Carey⁴ were the first to report using total hip arthroplasty (THA) to manage avascular necrosis of the femoral head in patients with Gaucher disease. In 1988, Goldblatt and colleagues⁵ reported excellent limb mobility and freedom from pain among 8 patients who underwent 15 THAs despite "resistance among colleagues to perform this operation among Gaucher's disease."

Hip arthrodesis in young patients⁶ is appropriate when the contralateral hip joint is preserved, as is (inexplicably) the case in many patients with Gaucher disease. Our experience (Lebel and colleagues⁷) with 36 THAs in both young and old patients receiving enzyme therapy and with various procedures and hardware showed almost uniformly excellent results with a relatively low rate of complications and

high levels of patient satisfaction and function. Other investigators have reported comparable results.^{8,9} Nevertheless, since 1992, cemented prostheses have been superseded for virtually all THAs performed at our center.

In this article, we describe the cases of 12 patients who had Gaucher disease, underwent cementless THA, and were followed for more than 7 years.

PATIENTS AND METHODS

In Israel between 1986 and 1997, 29 patients underwent 33 hip arthroplasties. Cementless prostheses were used in 19 of these cases, and 12 patients (15 hips) were followed up for more than 7 years. Demographic and Severity Score Index (SSI)¹⁰ data were obtained from patient charts.

For the study, Dr. Lebel performed complete physical examinations, collected perioperative and postoperative details from patients' medical files, and used the Harris Hip Score questionnaire to obtain functional data.^{11,12} Also reviewed were routine annual evaluations, including annual anteroposterior and lateral hip radiographs and other imaging as needed. Identification of implant failures was based on clinical evaluations and radiographic evidence of loosening; implant subsidence or malpositioning was recorded with the Gruen system.

Our institutional review board (Helsinki Committee) deemed that patient consent was not required for this retrospective analysis.

RESULTS

In all patients, avascular necrosis of the hip joint was related to Gaucher disease, and the indication for surgery was hip joint pain unrelieved by pain medications or other conservative measures. Table I lists the demographic and SSI data, and Table II lists THA data. Before THA, no patient had undergone any other surgical intervention for the joint.

Mean age at surgery was 53 years (men) and 32 years (women). Mean SSI was more than 15 points, implying moderate to severe disease.

The majority of surgeries were performed at our center. In all cases, the standard anterior (anterolateral) approach was used. In 11 THAs, the Aesculap hip prosthesis was used. As the bony structure in Gaucher disease is normal, proximal fixation was deemed appropriate. Most of the femoral stems (13/15 stems, 11/12 patients) were short, proximally press-fit, coated (hydroxyapatite or plasma-spray) stems. Acetabular implants were all press-fit, coated, metal-backed cups; reinforcement pelvic rings were used only in the 2 cases of revision of the acetabular component

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Table I. Demographic Data, All Patients

Patient	Sex	SSI	Genotype	Age at Surgery (y)	Enzyme Therapy	Spleen Status
1	M	21	1226/1226	36	No	Splenectomy
2	M	16	1226/84GG	58	No	Intact
3	M	16	1226/1226	44	Yes	Intact
4	M	17	1226/1226	64	Yes	Splenectomy
5	M	6	1226/431	50	Yes	Splenectomy
6	M	19	1226/84GG	51	Yes	Splenectomy
7	M	7	1226/1604	69	No	Intact
8	F	18	1226/84GG	41	Yes	Splenectomy
9	F	14	1226/1226	34	Yes	Intact
10	F	12	1226/84GG	31	Yes	Splenectomy
11	F	27	1226/IVS2+1	37	Yes	Splenectomy
12	F	9	1226/84GG	17	Yes	Intact

Abbreviation: SSI, Severity Score Index.

(to be described). Surgery time ranged from 2.5 to 3 hours.

There were no serious perioperative complications (ie, infections, pulmonary embolisms, major bleeding). Postoperative hemoglobin levels (data available for 7/12 patients) were 2.2 to 6.4 g/dL (mean, 4.6 g/dL) lower than preoperative levels.

Mean follow-up was 8.3 years (range, 7-18 years).

Partial or full weight-bearing was started immediately after surgery. All patients had a short, at-home rehabilitation and resumed independent ambulation within a few weeks.

Four patients required revision.

Patient 3, an active, relatively young man, complained of sudden hip pain 10 years after surgery and showed radiologic evidence of surface wearing.

Patient 8 (bilateral THA) sustained an implant dislocation that necessitated open reduction (closed reduction failed) on postoperative day 9. The acetabular component was malpositioned and was revised 11 months later. Eleven years after surgery (and after 2 deliveries), she complained of contralateral hip joint pain, which required revision of the pelvic component.

Patient 9 reported hip pain 6 years after surgery and showed radiologic evidence of acetabular loosening, which necessitated revision of the component (after 2 deliveries). Twelve months later, there was more evidence of loosening, also managed with a revision.

Patient 11 underwent revision 9 years after primary THA because of a huge infarcted area in the ilium (both surgeries were performed with local anesthesia because of pulmonary hypertension). The cup revision was accomplished with bone grafts only for filling; the stem was well integrated clinically and radiographically. As with all pathologic acetabular specimens, Gaucher cells were the predominant if not only type of cells present, regardless of years of enzyme replacement therapy, as was true of the other 3 revision cases.

Stem length was standard (ie, 180 mm from the center of rotation of the femoral head), and only mild changes were dictated by the diameter of the medullary canal. In most cases, the Aesculap prosthesis was used—the BiContact for the femur and the Plasmacup for the acetabular component (Aesculap AG, Tuttlingen, Germany).¹³ The Aesculap has been our preferred prosthesis for both Gaucher and non-Gaucher hip arthroplasties.

Stem length was measured with plain radiographs before surgery and with manufacturer instruments during surgery. With regard to ingrowth, standard radiographs were used to ascertain whether trabeculation occurred around the prosthesis. In 2 revision cases (patients 3 and 11), the stem was left in place (after attempts to dislodge it) and was found to have achieved excellent integration with the femur; in another case that required replacement (patient 9), excellent new bone growth was apparent around the proximal part of the stem.

The remaining patients did not complain of pain. Annual follow-up radiographs showed no signs of loosening of the implants that did not require revision. There were no cases of periprosthetic fracture. Although there was some degree of functional limitation related to the contralateral hip joint in 2 patients, neither patient was scheduled for revision of the operated hip or for primary THA of the contralateral hip.

Mean Harris Hip Score was 86.9 points. Most scores were in the good-to-excellent range (83-100 points). Only 3 patients had less than good scores, but there were extenuating circumstances in each case. Patient 2 had dementia and generalized low function, patient 8 had multiple sites of skeletal pathology, and patient 10 was pregnant when she was evaluated for this study. Patient 9 later suffered from ankylosis of the contralateral hip but refused surgical intervention and indicated she was satisfied with her current high-functioning status.

DISCUSSION

In this study of long-term follow-up of patients with Gaucher disease, a chronic disorder affecting the bony matrix, results with cementless primary THAs appeared to be superior to those we have obtained with cemented THAs in patients with this disease.⁷ Of 15 cemented hips (13 patients), 7 hips required revision within 5 to 10 years, and 1 other hip showed signs of loosening. Although patients were satisfied in both series, cementless THAs will prove to be an asset in revision cases. Prosthesis type may not be as critical as performing a cementless procedure.

Initially, surgeons used cemented hip implants for Gaucher disease because they assumed that osteoblast/osteoclast dysfunction would retard osteointegration. In addition, inflammatory mediators that are inimical to bone

Table II. Results of Total Hip Arthroplasty

Patient	Side	Revision	Follow-Up (y)	Harris Hip Score	Prosthesis
1	R, L	No, no	18, 18	95	Not known, not known
2	R, L	No, no	11, 17	76	Aesculap, Aesculap
3	L	Liner	12	97	Aesculap
4	R	No	10	95	Aesculap
5	L	No	7	99	Aesculap
6	L	No	7	100	Aesculap
7	R	No	7	97	Biomet
8	R, L	R (no), L (cup ×2)	13, 13	50	Aesculap, Aesculap
9	R	Cup ×2 twice	10	83	Aesculap
10	L	No	7	57	Landoz
11	L	Yes	7	94	Aesculap
12	R	No	7	100	Aesculap

Abbreviations: R, right; L, left.

are increased in Gaucher disease,^{14,15} increasing concern about integration of prostheses. There was also a suggestion that enzyme therapy may reduce trabecular bone.¹⁶ Pathologic evidence from our patients' cases, described here, showed densely packed Gaucher cells with and without enzyme therapy; nevertheless, radiologic evidence over time has shown good integration into the matrix.

The success of arthroplasties in this cohort may also be partially attributed to an integrated approach to preoperative protocols to assess coagulopathies unrelated to Gaucher disease, to provide prophylactic antibiotic cover especially in splenectomized patients, and to encourage early ambulation.

Although cementless arthroplasty may be equally applicable to nontraumatic osteonecrosis of the hip in patients with other metabolic disease, such as sickle cell disease,¹⁷ it may be more difficult to achieve because of nonsurgical concerns that increase morbidity¹⁸ and even mortality.¹⁹ However, when the etiology of osteonecrosis is nontraumatic, and hematologic and preoperative anti-infective measures are adequate, cementless hip arthroplasty has been used successfully.²⁰⁻²²

Surgeons may prefer cementless over cemented prostheses because of the technical facility and related advantages of cementless procedures in primary arthroplasty and in revisions, regardless of patient medical history. Nevertheless, the good results that we have reported in this article—in a model of bone matrix that progressively deteriorates or has a decreasing trabecular matrix—may have implications for indications for hip arthroplasty in other patients with metabolic bone disease.

AUTHORS' DISCLOSURE STATEMENT AND ACKNOWLEDGMENTS

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