Custom total hip arthroplasty in skeletal dysplasia

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Received: 27 May 2011 / Accepted: 20 June 2011 / Published online: 13 July 2011
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Abstract

Purpose Osteochondrodysplasias are characterised by aberrant cartilage and bone development; consequently, patients may be subject to premature hip degeneration. In this population hip arthroplasty outcomes are variable. This series reviews the use of custom femoral implants in total hip replacements for patients with severe skeletal dysplasia. **Methods** We reviewed 14 hips in nine patients with genetic dwarfism. Computed tomography (CT) scans of the pelvis and femurs were used to create custom titanium stems. Postoperative radiographs were reviewed for migration, osteolysis, fracture and restoration of leg length. Pre- and postoperative Harris hip scores (HHS) were compared, with a mean follow-up of 36.3 months. Mean age was 36.5 years (range 15–50 years) and mean height was 48 inches (range 42–55 inches). Three joints were classified as Crowe type I, two type II, three type III and six type IV.

Results Three hips (21%) required revision: two for stem migration and one for acetabular osteolysis. Eleven hips (79%) were stable with signs of osseointegration. No fractures, nerve damage or dislocations occurred. Two stems required cement fixation, and no liners were constrained. Significant osteolysis was seen on three radiographs. All patients walked independently, and mean HHS improved from 45 (24–58) to 71 (47–89). All leg length discrepancies were restored to within 3 mm of equal.

Conclusions This study reports good midterm outcomes in a variety of skeletal dysplasias, thus confirming the efficacy of custom femoral implants in the management of this patient group.

Introduction

Skeletal dysplasia encompasses a number of genetic conditions characterised by abnormal cartilage and bone development [1]. Though patients may be phenotypically similar, with short stature, hyperlordosis and coxa vara, the underlying genetic conditions that predispose to early joint destruction are markedly different [2]. A number of congenital osteochondrodysplasias are characterised by arrested epiphyseal development, resulting in delayed endochondral ossification, and thus an unsupported cartilaginous surface, ultimately leading to the obliteration of the loaded joint [3]. The clinical and functional problems encountered by this patient group become apparent in early adulthood, whereby femoral head, neck and acetabular pathology create an incongruous restricted hip joint, compounded by accompanying flexion contractures [3, 4] (Fig. 1). Consequently, gait disturbance becomes markedly apparent in adolescence, while concurrent foot and knee deformities may further alter the lower limb mechanical axis. As a result of such distorted anthropometry early surgical intervention is commonly proposed, whereby femoral and acetabular osteotomies are frequently used as means of hip preservation [5]. Yet in many patients these interventions are purely temporising, thus necessitating a more definitive procedure. The use of early arthroplasty in severely dysplastic hips primarily relied on cement fixation of both components [6, 7], while recent mid- and long-term results have been published for cementless press fit implants [5, 8, 9]. The variable outcomes reported are mainly due to the challenges particular to skeletal dysplasia, where severely altered acetabular anatomy is compounded by short bowed femurs. Previous studies have reviewed the use of custom and non-custom prosthetics for both femur and acetabulum [5, 10–12],

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Fig. 1 Preoperative patients with pseudoachondroplasia and Hurler's syndrome

yet an unusually high incidence of periprosthetic femoral fractures and nerve injuries were noted [5, 9, 12]. Moreover, studies that advocate the sole use of custom prostheses have highlighted the necessity for cement fixation of femoral components [5], due to deformity and concomitant osteoporosis. With this variability in outcomes, the objective of this study is to review the midterm survival of customised femoral components in the management of hip osteoarthritis in skeletal dysplasias.

Materials and methods

Between 1996 and 2008, 14 primary total hip arthroplasties were performed on nine patients with skeletal dysplasia—three male and four female. Average age at time of surgery was 36.5 years (range 15–50 years), average height 48 inches (42–55 inches), weight 109 lb (250–74 lb) and

average time to follow-up 36.3 months (eight–121 months) (Table 1). All diagnoses were confirmed by genetic testing and/or radiographic assessment, and all operations performed by two senior surgeons (MF and MB). Of the 14 hips, nine had undergone previous surgical procedures (six pelvic acetabular osteotomies, three Harrington rods). Three joints were classified as Crowe type I, two type II, three type III and six Crowe type IV.

Operative technique

The posterolateral approach with the patient in the lateral decubitus position was used in all operations. In three cases a portion of the gluteus maximus tendon was reflected to assist exposure. Femurs were initially hand reamed, and in some cases reaming was completed with intramedullary shoulder set reamers; the sclerotic most proximal femoral bone was milled using a Midas burr, before a custom broach

Table 1 Patient demographics

Patient no.	Age	Side	Diagnosis	Previous hip surgery	Crowe score	Implant manufacturer
1	47	R	Pseudoachondroplasia	Femoral osteotomies	2	Biomet
1	47	L	Pseudoachondroplasia	Femoral osteotomies	1	Biomet
2	43	R	Diastrophic dysplasia	Nil	3	Biomet
2	43	L	Diastrophic dysplasia	Nil	3	Biomet
3	34	R	Multiple epiphyseal dysplasia	Femoral osteotomies	3	Biomet
3	34	L	Multiple epiphyseal dysplasia	Femoral osteotomies	4	Biomet
4	25	L	Multiple epiphyseal dysplasia	Nil	1	Biomet
4	24	R	Multiple epiphyseal dysplasia	Nil	1	Biomet
5	44	R	Diastrophic dysplasia	Femoral osteotomies	4	Biomet
5	50	L	Diastrophic dysplasia	Femoral osteotomies	4	Biomet
6	15	L	Hurler's syndrome	Femoral osteotomies	4	Omnifit
7	32	L	Achondroplasia	Nil	4	Biomet
8	35	R	Diastrophic dysplasia	Nil	4	Biomet
9	38	L	Hurler's syndrome	Nil	2	Zimmer

was used for trial impaction prior to the custom implant being used for trial reduction. In three cases, the decision to use cement was taken preoperatively following radiographs and/or bone scans; in this setting cement was passed into the canal in a retrograde fashion with the cement gun, and finger impacted due to the small size of the femoral canals. The custom stem was then pressed into position. All acetabular components were press fit, with off the shelf sequential reamers used in all cases; typically, cups were medialised to obtain sufficient coverage and assist in the reduction of joint load [13]. Preoperative prophylactic antibiotics were used in all cases. No added precautions apart from standard total hip precautions were imposed postoperatively with all patients discharged weight-bearing as tolerated.

Custom design

A preoperative computed tomography (CT) scan of the proximal femur and knee is used to determine the shape, head centre location and anteversion of the femoral component. The proximal body of the implant is designed to transfer load in a corkscrew fashion starting proximally in the posteromedial region of the femur and rotating to the anterolateral region more distally. This design of bone-implant contact is intended to resist the posteriorly directed load on the femur. To resist bending loads, the stem is extended distally as a smooth generally straight cylinder. Generally the anteversion of the native femur is excessive and if the implant were designed to match the original head centre, there would be risk for anterior dislocation. Instead, the anteversion of the component is reduced to a more normal value (typically between 20 and 25°). Head height is measured on the anteroposterior (AP) radiograph and templated with an image of the device. Head height is generally matched to the native head height. Once the design is approved, the implant is manufactured from titanium alloy. Acetabular component sizes were also assessed digitally from CT scans due to typically superior accuracy [14].

Radiographic and functional assessment

Follow-up radiographs were reviewed for evidence of osteolysis and component migration, whereby definite failure was defined as migration greater than 3 mm, and impending failure as circumferential radiolucency greater than 2 mm at the bone-metal interface. Evidence of osseointegration was assessed as was incidence of heterotopic ossification as defined by Brooker et al. [12]. Preoperative and postoperative films were compared for femoral neck-shaft angle, offset and leg length, while initial Harris hip scores were noted at the preoperative visit and then compared to scores from the most recent follow-up.

Statistical analysis was performed with paired *t* tests using SigmaStat and significance set at $p < 0.05$.

Results

Radiographic analysis

At the most recent follow-up, 11 of 14 (79%) hips remained unrevised. All acetabular shells were press fit, additional screw fixation was used on 12 acetabular components, though no acetabulae required bone grafting. Seven hips required elevated liners but no constrained liners were used; average acetabular shell size was 44.2 mm (40–56 mm), and average head size was 30 mm (28–32 mm). Short external rotator tenotomy was required in one hip due to flexion contracture of 50°. Average femoral offset increased from 20.7 mm (18–24 mm, ± 2.4) to 37 mm (33–41 mm, ± 3.1), and one femur required additional graft due to bone loss from previous surgical treatment. Of the six hips with leg length discrepancies, all lengths were restored to within 2 mm of equal. Three hips required revision, and one acetabular component was revised to a larger shell three weeks postoperatively due to migration; the same patient developed a *Staphylococcus aureus* infection two months postoperatively. One patient required revision eight years following the index procedure due to aseptic loosening of the cemented femoral component, though this was in the setting of concomitant new onset morbid obesity. The third patient underwent revision of a cemented stem due to migration at four years. This subsequently required cementless re-revision again for aseptic loosening 12 months later; the third custom component was well fixed. No fractures, dislocations or nerve injuries occurred. The remaining 11 hips showed signs of osseointegration, though three were found to have asymptomatic radiographic evidence of heterotopic ossification—all grade II. Of these 11 hips, evidence of osteolysis was noted in three radiographs, in Gruen [15] zones 1, 4 and 7.

Functional analysis

There was significant change in Harris hip scores postoperatively with a mean increase from 44.8 (24–58) to 71.8 (47–89). All ranges of motion showed a degree of improvement, though none were statistically significant. The presence of coexisting vertebral disease affected the scores in relation to limping, yet a notable improvement was apparent from the mean change. The most marked improvement was noted in pain scores with 78% of patients reporting hips as pain free at last follow-up; in addition, the climbing of stairs showed a significant enhancement in 86% of hips. Seven patients mobilised independently, one used a walker outdoors, though this was similar to

preoperative ambulation, while one patient used a cane full time. The characteristic waddling gait was still present in all patients, though 86% of hips subjectively performed “better than satisfactory”.

Discussion

Patients with skeletal dysplasias are prone to complex deformities, leading to the early onset and often debilitating osteoarthritis of the hip joint. The goal of all total hip arthroplasty procedures is primarily to relieve pain and secondarily to restore a biomechanically functional joint; within this patient subgroup this is partially achieved through the resolution of leg length discrepancies, but also in the restoration of functional anatomy in the setting of spinal and lower limb deformity. The altered biomechanics of the dysplastic hip is compounded by the exaggerated lumbar lordosis, thus altering the load and structural properties of the hip joint, factors considered at custom component creation. In previous series femoral deformity was thought to be a major influence on the number of perioperative and postoperative periprosthetic femoral fractures. Chiavetta et al. [9] noted a near fourfold increase in the number of postoperative femoral fractures, while earlier studies [5, 6, 10, 12, 16] also noted an increased rate, predominantly occurring at the tip of the prosthesis or cement mantle. With the move away from cemented stems, the recent Lim et al. [8] study illustrated good results with the press fit modular S-ROM femoral prosthesis, with only one reported femoral fracture at insertion. They suggested that the modular nature of the component allowed for greater intraoperative variation and a more accurately achieved femoral offset, thus lowering the femoral fracture rate. Our series compares favourably with these data; no fractures occurred in any procedure, yet component alignment and offset remained satisfactory. In addition,

previous reviews of custom components in this patient group stressed the absolute necessity for cementing of stems due to canal size and poor bone stock [5]. Conversely, we found that although low bone density stipulated cementing components in three hips, in the 12 remaining hips we were able to use press fit prosthetics. The use of CT created components facilitates more accurate measurements of intramedullary diameter, offset, version and neck shaft angle; this is in contrast to custom prostheses created in the Huo et al. [11] series based on plain radiographs. In addition, the close working relationship between surgeon and biomechanic resulted in a porous component that can be fixed in the narrowest of canals; moreover, the enhanced stability attributable to press fit stems [17] is pivotal in this population where load is unevenly distributed across the pelvis. This premise was clearly evident in our series where two of the three cemented components required revision for aseptic loosening.

Results from this study support the continual use of the posterolateral approach without modification. Previous series [8, 12] have advocated the use of a direct lateral or anterior approach to facilitate capsule release, as well as highlighting the absolute need for tenotomies of the sartorius, rectus femoris and/or the tensor fascia lata. Our lack of deviation from typical practice is in part permitted through use of custom prosthetics, whereby many measurements are determined at stem creation, and though the most severe of contractures required tenotomy, capsular release was seldom required for exposure. The high historical incidence of nerve injuries may in part be due to the tendency for widespread soft tissue release and difficulties gaining exposure, yet our series saw no transient or permanent nerve damage whilst maintaining the standard approach.

The radiographic outcomes achieved with the use of custom prostheses proved favourable in our study, yet it is the marked functional improvements relating to daily activities which showed the most promise. The distinct

Fig. 2 Postoperative patients with pseudoachondroplasia and Hurler’s syndrome

reduction in pain and the increased ease with which patients managed stairs were subjectively the most important outcomes; furthermore, these improvements particularly in stair climbing and the use of public transport were significant in the light of patient stature.

Ultimately, for any complex arthroplasty especially those involving custom implants, the need for exacting preoperative planning is essential; a number of patients had undergone previous operations which further altered pelvic anatomy. Through CT and multiple radiographic views, the absolute anatomy of the complex pelvis can be assessed, as well as the early determination of cortical thickness and acetabular depth. Measurements discernible from a CT allow for the creation of a monoblock custom prosthesis that not only will better conform to the deformed femur but also compensate for the often dysplastic acetabulum, thus providing a platform for solid bony ingrowth and improved joint functionality (Fig. 2). Furthermore, this degree of planning does not end in the operating room, but instead extends to an often prolonged period of rehabilitation. Though the patients in this group are often younger than the archetypal arthroplasty patient, they are burdened with multiple joint diseases and altered anthropometry, thus requiring prolonged and intensive functionally specific therapy. This study reaffirms total hip arthroplasty as a good solution to hip osteoarthritis in skeletal dysplasia; furthermore, we illustrate the effectiveness of maintaining the posterior approach and uncemented components. Though small in number, this study adds to the limited body of work investigating the sole use of custom prostheses in skeletal dysplasia. The biggest perceived limitation or disadvantage associated with custom use is the seemingly prohibitive costs of implants amounting to approximately US \$5000 per complete total hip prosthesis [18]; many feel this cost and the four to five week period needed for modelling inhibit its use as a primary option in any patient group. Yet, this cost is offset by the superior results garnered both radiologically and functionally. Ultimately, the use of total hip arthroplasty in patients with severe skeletal dysplasias has long been known to be of value; this series illustrates the efficacy of custom femoral components as a definitive management option in this diverse patient group.

Conflict of interest The authors declare that they have no conflict of interest.

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