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Case Series

Long Term Outcomes and Survivorship of Bilateral Lower Limb Arthroplasties in Patients with Multiple Epiphyseal Dysplasia

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Abstract: Background: Multiple epiphyseal dysplasia (MED) a rare form of skeletal dysplasia with early-onset osteoarthritis affecting the hip and knee joints. There is paucity of evidence regarding the long term outcomes of arthroplasty in this patient cohort. This is the first study to describe the outcomes of bilateral arthroplasty of the hip and knee in patients with MED. **Methods:** A prospective study of patients with MED who underwent bilateral staged total hip arthroplasty (THA) and bilateral simultaneous total knee arthroplasty (TKA) between July 2013 to April 2014. Preoperative and postoperative clinical and radiographic assessment including functional outcomes namely Merle d'aubigne score, Harris hip score (HHS), Knee society score (KSS) and numeric rating scale (NRS) for pain were recorded in prospective database and evaluated. **Results:** 3 patients (female - 1 / male - 2) with MED who underwent bilateral lower limb arthroplasty of the hip and knee were reviewed periodically with an average followup of 7.31 years. Mean preoperative Merle d'Aubigné score (6.33 ± 0.58), HHS (31.83 ± 5.31) and KSS (28.67 ± 7.97) increased following bilateral lower limb arthroplasty with mean postoperative Merle d'Aubigné score (16.33 ± 0.58), HHS (84.33 ± 5.99) and KSS (86.33 ± 7.09) (P<0.001). Mean NRS decreased from 7.33 ± 0.58 in the preoperative period to 0.67 ± 0.58 at the most recent followup (P<0.001). One patient had heterotrophic ossification following THA. There were no revisions. **Conclusions:** Bilateral lower limb arthroplasty of the hip and knee is a good treatment option to restore function and mobility in MED patients with advanced degenerative arthritis. Careful preoperative planning, meticulous surgical techniques, patient focussed postoperative rehabilitation with multidisciplinary team approach are vital to ensure good functional outcomes and implant survivorship.

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1. Introduction

Multiple epiphyseal dysplasia (MED) or Fairbank's disease is a rare form of skeletal dysplasia which severely affects the articular cartilage resulting in severe deformities [1]. It is genotypically and phenotypically heterogenous, and the inheritance can either be autosomal dominant with variable penetrance or autosomal recessive [2]. It presents between 5-14 years of age, with an incidence of 1:10000 [2, 3]. Autosomal dominant case mutations of the collagen oligomeric matrix protein (COMP), collagen type IX α-1, collagen type IX α-2, collagen type IX α-3, and matrilin-3 genes have been reported in patients with MED [2, 4]. The autosomal recessive variant is caused by a mutation of the sulphate transporter gene SLC26A22 [2]. Additionally, there can be variants that are sporadic in origin [5]. Patients typically present in late childhood. However, some do not show any symptoms till adulthood and present with predominant affliction of the hip and knee joints [2, 3, 5]. Spinal involvement is relatively uncommon and manifests as scoliosis [2, 5].

Clinical symptoms may include pain, joint stiffness, waddling gait, and limb length variation depending on the severity and type of joint involvement [1-3, 5]. Hip joint involvement leads to an incapacitating early onset degenerative disease which often begins between the second and fourth decade of life [6-8]. Total hip arthroplasty (THA) has been reported in the literature in the treatment of advanced hip degeneration [6, 8, 9]. In patients with involvement of the knee joint, total knee arthroplasty (TKA) coupled with acute deformity correction as required has been described as a part of surgical management [10]. Furthermore, with combined hip and knee joint involvement, arthroplasty of both the aforementioned joints is essential to restore the functional activity [11].

Arthroplasty remains the treatment modality of choice in majority of these patients [2, 7]. However, the distorted anatomy of the afflicted joints makes the surgery challenging [10, 11]. Patient factors such as poor bone quality, younger age, along with surgical factors such as distorted anatomy which may necessitate the use of customized prosthesis may contribute to a higher morbidity and mortality [11]. Some studies have reported THA and TKA as a definitive treatment modality in these young patients [6, 7, 10, 11]. However, there is paucity of literature regarding the optimal orthopaedic management using THA and TKA in patients with MED and their long term outcomes [8]. Furthermore, to the best of our knowledge, there is no study in the current literature describing the long-term outcomes of combined THA (staged) and TKA (simultaneous) in patients with MED. Hence the primary aim of our study is to evaluate the long-term functional and radiological outcomes, including complications and survivorship of combined THA and TKA in patients with MED. The secondary aim is the highlight the technical challenges encountered during arthroplasty in these patients and describe some novel solutions to overcome them.

2. Materials and Methods

2.1. Study Design and Participants

This prospective study was undertaken at Sparsh Hospital, Infantry road, Bangalore, India. We describe three patients diagnosed with MED who underwent bilateral hip and knee arthroplasties between July 2013 – April 2014. Institutional review board approval was obtained, and all patients signed a written consent before participating in the study.

All the three patients had previously undergone genetic analysis, which confirmed the diagnosis of MED. All patients attended preoperative assessment during which routine parameters including demographic details, Body mass index (BMI) and Charlson comorbidity index (CCI) were recorded in the prospective arthroplasty database of the institution.

Demographic details, patient characteristics and preoperative clinical parameters were recorded (Table 1 and Table 2). Preoperative radiographs included pelvis, bilateral hips, bilateral knee and scanograms to assess limb alignment (Figure 1). All the surgical procedures were performed by the two senior authors (SSP and RM). TKAs were performed simultaneously, while THAs were performed in a staged manner. Surgical details of the three cases are presented below.

Table 1. Demographic details, preoperative parameters and follow up duration of the MED patients.

Parameter	Case 1	Case 2	Case 3
Age	46	19	36
Gender	Male	Male	Female
Height (m)	1.44	1.43	1.35
Weight (kg)	55.99	50.10	41.92
BMI (kg/m^2)	27	24.5	23
Charlson comorbidity index	1	1	1
Numeric rating scale (pain)	7	8	7
Merle d'Aubigné score	6	7	6
Followup duration (years)	7.07	7.23	7.75

Table 2. Preoperative and postoperative clinical parameters of lower limb joints and functional outcome scores.

	Case 1		Case 2		Case 3	
	Preoperative	Postoperative‡	Preoperative	Postoperative‡	Preoperative	Postoperative‡
Range of motion (ROM) - degrees						
Hip† – right flexion	30 – 40	0 - 90	20 – 50	0 - 100	15 – 50	5 - 85
Hip† – left flexion	30 – 40	0 - 90	30 – 80	0 - 100	20 – 60	0 - 80
Knee – right	0 – 15 – 70	0 – 10 – 110	0 – 25 – 60	0 – 5 – 100	0 – 15 – 80	0 – 5 – 90
Knee – left	0 – 10 – 70	0 – 10 – 110	0 – 30 – 60	0 – 5 – 100	0 – 20 – 70	0 – 10 – 100
Hip – right	30	0	20	0	15	5
Hip – left	30	0	20	0	20	0
Knee – right						
coronal*	-20	0	-20	0	-15	0
sagittal	15	10	25	5	15	5
Knee – left						
coronal*	-20	0	-20	0	-15	0
sagittal	10	10	30	5	20	10
Harris Hip Score						
right	26	88	37	91	34	85
left	26	88	38	77	30	77
Knee Society Score						
right	32	90	18	80	34	94
left	30	80	20	80	38	94
Merle d'Aubigné score	6	17	7	16	6	16
Numeric rating scale	7	1	8	1	7	0

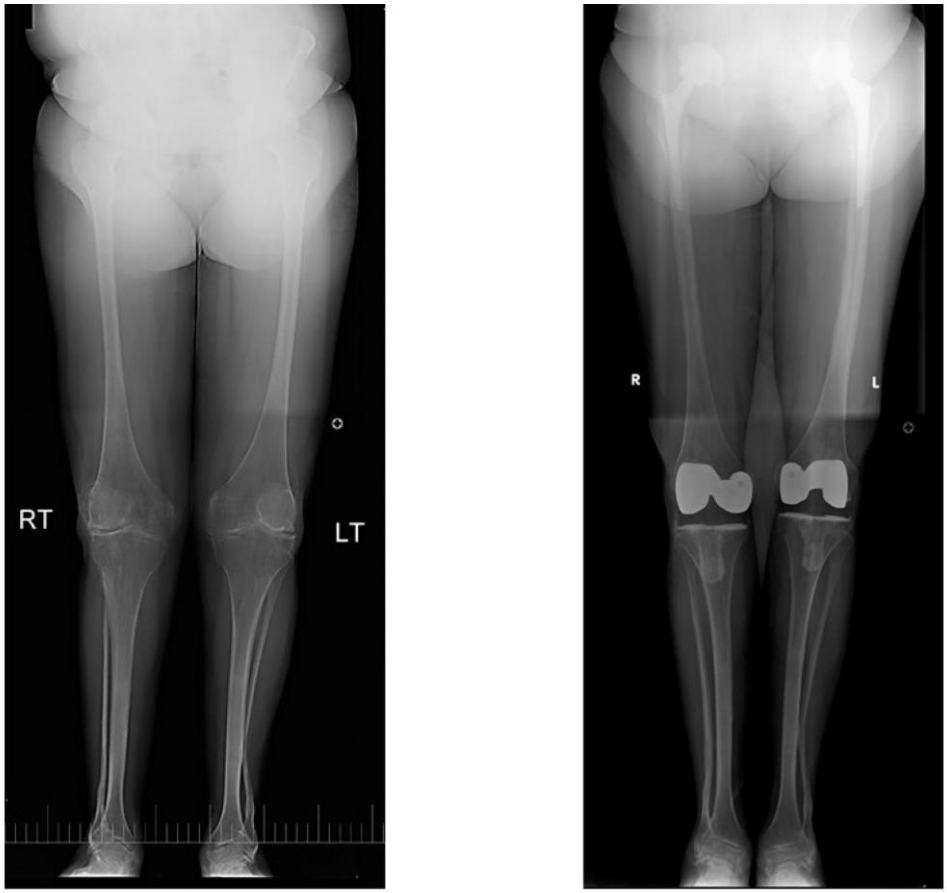

Figure 1. Preoperative (a) and postoperative (b) lower limb scanograms.

2.2. Surgical Procedure

2.2.1. THA

All the six THA were performed in a staged manner (few days apart) using a modified Hardinge's approach and an uncemented hydroxyapatite coated femoral stem, uncemented acetabular cup with metal on polyethylene or ceramic on ceramic bearing couple were implanted. DePuy implants (DePuy, Warsaw, Indiana, USA) were used (Table 4). Wound closure and application of non-adherent wound dressing and circumferential bandage were performed in a routine manner.

Table 3. Improvement in functional outcomes following bilateral lower limb arthroplasty.

Outcome score	Preoperative (Mean ± SD)	Postoperative (Mean ± SD)	*P value*
Harris Hip Score	31.83 ± 5.31	84.33 ± 5.99	<0.001
Knee Society Score	28.67 ± 7.97	86.33 ± 7.09	<0.001
Merle d'Aubigné score	6.33 ± 0.58	16.33 ± 0.58	<0.001
Numeric rating scale (pain)	7.33 ± 0.58	0.67 ± 0.58	<0.001

Table 4. Type and sizes of implants used during hip and knee arthroplasty in MED patients.

Implant	Case 1	Case 2	Case 3
Total hip arthroplasty Right			
Acetabulum cup (mm)	50	58	50
Liner material	Poly	Poly	Poly
Femoral head size (mm)	28[†]	32[*]	28[*]
Femoral head offset (mm)	+1.5	+9	+1.5
Femoral stem size	6	14	6
Left			
Acetabulum cup (mm)	54	60	48
Liner material	Ceramic	Poly	Poly
Femoral head size (mm)	36[†]	32[*]	28[*]
Femoral head offset (mm)	+1.5	+9	+1.5
Femoral stem size	8	13	6
Total knee arthroplasty Right			
Femur	2	4	1.5
All poly tibia size	2	4	1.5
All poly tibia thickness (mm)	12.5	8	10
Left			
Femur	2	5	1.5
All poly tibia size	2.5	4	1.5
All poly tibia thickness (mm)	8	12.5	10

2.2.2. TKA

All patients underwent bilateral simultaneous TKA procedures under spinal anaesthesia using routine midline incision and medial parapatellar approach. During TKA a posterior-stabilised cobalt-chromium femoral component and an all-poly tibial component were implanted using cement without patella resurfacing. Depuy PFC sigma implants (DePuy, Warsaw, Indiana, USA) were used (Table 4). Wound closure and application of non-adherent wound dressing and circumferential bandage were performed in a routine manner.

2.3. Technical challenges and novel solutions used during surgical procedures

2.3.1. Case 1

THA: In the right hip, it was noted that the greater trochanter was hinging on the ilium couple with soft tissue contracture involving the iliotibial (IT) band and iliopsoas complex thereby making the surgical exposure difficult. Safe dislocation of the femoral head was not possible in both the hips as the femoral head was deformed and deeply seated within the acetabulum. Additionally, the femoral neck was noted to be short and broadened thereby making the osteotomy challenging. Hence, an in-situ osteotomy of the femoral neck was done. Following preparation of the acetabular socket with sequential reaming technique, a 50 mm acetabular cup was implanted. On the femoral side, the narrow canal was located with 2.5mm drill bit and pencil reamer. Subsequently, a guide wire was passed into the femoral canal under fluoroscopic guidance. After confirmation of the satisfactory position of the guidewire, flexible reamers were used in the initial preparation of the femoral canal. This was followed by sequential broaching to prepare the femur for stem implantation. The narrow canal compelled the usage of a size 6 stem (Table 4). Iliopsoas complex, IT band and adductor release was necessary to achieve reduction. In the

left hip, similar intraoperative findings were present which necessitated the above surgical steps. Postoperatively, lower limb lengthening of 3 cm was noted bilaterally with no demonstrable sciatic nerve palsy.

TKA: Medial parapatellar arthrotomy was used for the exposure during knee arthroplasty. Eversion of the patella was challenging as it was low-lying. Osteophytectomy of the femur, tibia was performed. In both the knees, distal femoral and proximal tibial condyles were enlarged, deformed with a very narrow canal, and altered bony landmarks. This resulted in a relative anteroposterior to mediolateral dimension mismatch. Hence, a combination of extraarticular and intraarticular jigs was used as a reference prior to undertaking femoral resection. Precautions were taken not to breach the cortex while inserting the intramedullary jig and fluoroscopic guidance was used to confirm the position. Posterior osteophytes in the distal femur were excised with curved osteotomes and capsular release was performed to achieve complete knee extension. An additional 2mm proximal tibial resection was necessary in the right knee to achieve balanced flexion and extension gaps.

2.3.2. Case 2

THA: Bilateral dysplastic acetabulum with broadened and distorted femoral head were present. The femoral head could be safely dislocated out of the acetabulum. Femoral neck was noted to be broad and short. Subsequently, sequential reaming of the deformed acetabulum was performed carefully to create a socket shape and a size 58 mm cup was placed in the right hip (Table 4). Careful femoral preparation under fluoroscopic guidance was performed as described in the above case. The femoral endosteal canal was noted to be deformed but was capacious and accommodated relatively larger femoral stem sizes compared to case 1 (Table 4). Postoperatively, lower limb lengthening of 2.7 cm was achieved with stable hip movements.

TKA: Similar extensile surgical exposure using a medial parapatellar approach was performed as in case 1. A hyperplastic patella (megapatella) was present bilaterally. This necessitated debulking and clearance of osteophytes to enable safe eversion of the patellae. The distal femoral condyles and the proximal tibial condyles were hyperplastic with rotational deformities. Hence a combination of transepicondylar axis (TEA), Whiteside's line and the posterior femoral condyle resections were performed perpendicular to the proximal tibial resection. The tibial baseplate placement was lateralized to overcome the extraarticular varus of the tibial shaft and achieve optimum coverage and avoid overhang. The overall component sizes were relatively larger compared to case 1 (Table 4).

2.3.3. Case 3

THA: Extensile surgical exposure was performed using modified Hardinge's approach. The acetabulum was relatively well formed. The femoral head was safely dislocated out of the acetabulum, with no neck shortening. Acetabular and femoral preparation were performed as described in the previous cases. Overall, the component sizes were similar to case 1 with the left hip accommodating only a 48mm acetabular cup (Table 4).

TKA: During the bilateral TKA, extensile surgical exposure using medial parapatellar approach were performed. Femoral and tibial deformities were similar to case 1. However, the patellae were found to be maltracking due to hypoplastic trochlea and lateral condyle. Hence, lateral soft tissue release was performed. Additionally, careful assessment of femoral component rotation was done using multiple reference methods as above and femoral resections were performed accordingly. A combination of soft and sclerotic bone was encountered in the femoral condyles. Screw augmentation was performed to ensure condylar stability at the transition area (Figure 2). Subsequently, an extra 2mm of proximal tibial resection was undertaken to achieve balanced flexion and extension gaps. Overall, the bony dimensions were small compared to the first two cases and implantation of the smallest femoral and tibial component sizes was performed (Table 4).

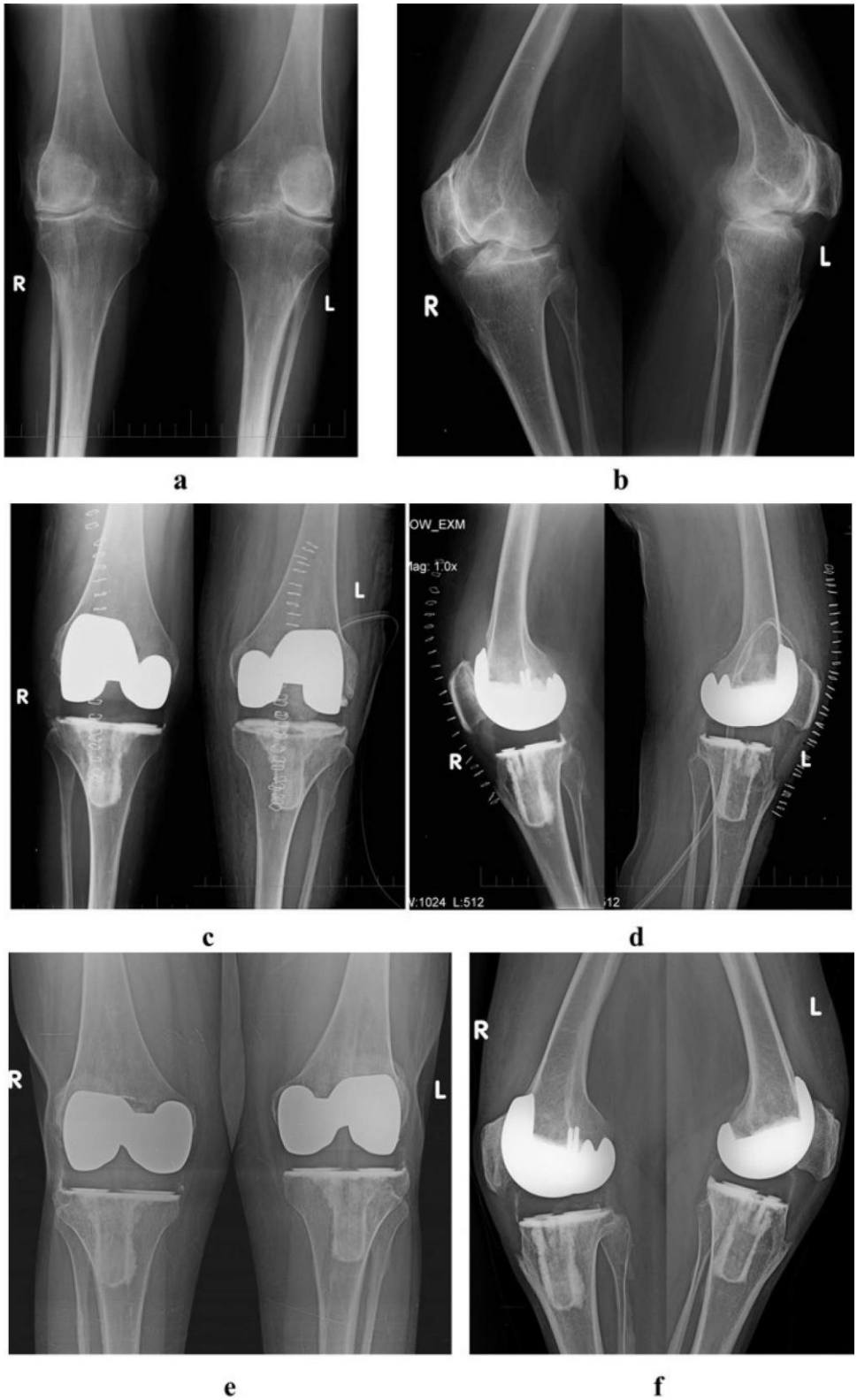

Figure 2. Bilateral knee radiographs - preoperative anteroposterior (a) and lateral (b), postoperative anteroposterior (c) and lateral (d), most recent followup anteroposterior (e) and lateral (f) views.

2.4. Postoperative care and rehabilitation

Postoperative pain relief included intravenous paracetamol 1 g six hourly, tablet tramadol 50 mg oral if required and injection diclofenac 75 mg twice daily if required. All patients received routine postoperative rehabilitation with supervised physiotherapy exercises. Weight bearing mobilisation as tolerated by the patient was commenced on the first postoperative day with the aid of a walking frame. Subsequently, the patients graduated from the initial walking frame to a pair of walking sticks and eventually to no aids in a phased manner during the first six to eight postoperative weeks.

2.5. Followup and Functional Outcome Scores

All the patients were evaluated for functional outcomes using Merle d'aubigne score [12], Harris hip score [13], Knee Society score [14] and numeric rating scale for pain [15].

Patients were followed up at 1, 3, 6, 12 months during the first year and annually thereafter. During the followup visits, patients were evaluated clinically and radiologically for functional outcomes and numeric rating scale for pain. Postoperative complications were recorded in the prospective database of the institution and implant survival was assessed accordingly.

2.6. Statistical Analysis

Statistical analysis was performed using SPSS, version 20, software (IBM, Armonk, NY). Descriptive statistics are presented as means with standard deviations for continuous variables such as mean patient-reported scores and counts or percentages for categorical variables. Means were compared between preoperative and postoperative outcome scores with paired t-test.

3. Results

3.1. Patient Characteristics

3 patients (female - 1 / male - 2) underwent bilateral primary THA (staged) and TKA (simultaneous) between July 2013 to April 2014. Patient demographics, clinical variables and followup duration are presented below (Table 1).

3.2. Clinical Parameters

3.2.1. Numeric rating scale (pain)

All the three patients reported a high pain score (range 7-8) prior to their lower limb arthroplasties. However, following the arthroplasty procedures their pain score improved and at the most recent followup the patients reported no significant pain (range 0-1) in their hip or knee joints (Table 2).

3.2.2. Range of motion – hip joints

All the three patients were noted to have fixed flexion deformity (range 15 to 30 degrees) in their hip joints prior to THA. Hip joint flexion was considerably restricted prior to surgery (range 40 to 80 degrees). However, post THA all the patients had improvement in their hip range of motion with correction of their fixed flexion deformity (Table 2).

3.2.3. Range of motion – knee joints

Knee joint assessment before TKA revealed fixed deformities in the coronal plane (range 15 to 20 degrees of varus) and sagittal plane (range 10 to 30 degrees) (Table 2). Scanogram radiographs for lower limb alignment confirmed the fixed deformity to be intraarticular. Range of motion in the knee joints was restricted prior to TKA with maximum flexion ranging between 60 to 80 degrees (Table 2). Following TKA, the lower limb alignment was corrected to neutral in the coronal plane and was maintained at the most recent followup. In the sagittal plane, neutral alignment was achieved immediately following

TKA. However, this was not sustained, and all the three patients were noted to have a fixed flexion deformity (range 5 to 10 degrees) at the most recent followup (Table 2).

3.3. Functional outcome scores

3.3.1. Merle d'Aubigné score

All the three patients had poor hip function prior to THA and reported low Merle d'Aubigné scores (range 6-7). At the most recent followup after the bilateral staged THA, the Merle d'Aubigné scores ranged between 16 to 17 representing an over improvement in the hip function of the three patients ($P < 0.001$) (Table 2 and Table 3).

3.3.2. Harris hip score (HHS)

Bilateral hip function was noted to be poor in the three MED patients in the preoperative assessment. After undergoing the bilateral THA, all the three patients had improvement in hip function. At the most recent followup, a significant improvement in HHS ($P < 0.001$) was noted in all the three patients ranging from 77 to 91 (Table 2 and Table 3).

3.3.3. Knee society score (KSS)

A combination of deformity and pain resulted in poor function with low preoperative KSS (range 18-38). Following bilateral simultaneous TKA, the joint deformity was corrected, and an increased range of motion was noted in all the three patients. This contributed to a significant improvement in the postoperative KSS and at the most recent followup, the KSS was high in all the three patients ranging from 80 to 94 (Table 2 and Table 3).

3.4. Complications

3.4.1. THA

All the three patients exhibited delayed wound healing (3 weeks) with serous discharge of their THA wounds. One patient (case 2) developed serosanguinous discharge from the left THA wound. Microbiological analysis revealed no growth. Subsequently, this patient underwent wound debridement and washout in the operation theatre. Following this additional procedure, the wound healed uneventfully.

Heterotrophic ossification (HO) was noted in the left hip during the postoperative followup in the above patient [**Figure 3**]. However, this condition remained largely asymptomatic and no further intervention was required.

Figure 1. Pelvis radiograph demonstrating heterotrophic ossification following left total hip arthroplasty.

3.4.2. TKA

In comparison, the TKA wounds in all the three patients healed uneventfully with primary healing during the first two postoperative weeks.

4. Discussion

Multiple epiphyseal dysplasia (MED) is a relatively rare genetic disorder. However, the orthopaedic manifestations of this condition have the potential to cause serious impact on the quality of life of the affected patients at a very young age [2, 3, 5]. Early and progressive degenerative arthritis sets in due to the incongruency and malformed cartilage. Arthroplasty has been described as treatment in these patients with advanced degenerative changes causing limitations in daily functional activities [11]. However, several patient and surgical factors such as young age, high functional demand, technical difficulties associated with arthroplasty, limited data regarding the long term survival of prosthesis

and the potential for revision surgery need careful consideration before undertaking arthroplasty [2, 11]. In comparison to the limited studies evaluating THA in MED patients, there is no study in the current literature reporting the long term outcomes of TKA [2]. The results from our study addresses this lacuna with demonstration of good long term outcomes and survivorship of bilateral THA and TKA in patients with MED.

In our study, the age of the patients ranged from 19 to 46 years which is comparable to other studies in the literature [6, 8]. Deformed anatomy common in all three cases were distorted acetabulum and femoral head, narrow canal, short femoral neck and upriding of the greater trochanter which was similar to previously published studies [6, 9]. In the knees, the femoral and tibial condyles were malformed with rotational deformities and altered landmarks. Furthermore, the patellae were relatively large, low lying and maltracking. However, there are no published studies to corroborate the various knee findings.

We performed bilateral lower limb arthroplasties including simultaneous TKA and staged THA following preoperative optimisation and planning with multidisciplinary team. The rationale for bilateral procedures included – 1) severe involvement of all four joints affecting daily functional activities and mobility 2) relative physiological fitness of patients to undergo bilateral arthroplasty 4) patient choice to regain independence and pursue career 2) treatment option which helps restore early overall function in young patients. Selection of implant sizes and bearing surfaces was undertaken based on careful preoperative templating and surgical factors as highlighted above. We noted improvement in patient reported outcomes following the bilateral arthroplasty procedures. Furthermore, the good function has been sustained as reported during the long term followup review (Table 2 and 3) (Figure 1 and 2). The bilateral THA and TKA enabled this cohort of young patients with MED to regain functional independence, pursue professional career of their choice and provide livelihood for their families.

The current literature has some studies reporting the outcomes of THA in patients with skeletal dysplasia including MED [6-9, 11, 16]. However, no study has reported the outcomes of bilateral arthroplasties (staged THA and simultaneous TKA) in patients with MED. Pavone et al. reported the results of bilateral THA in seven MED patients. They observed the Merle d'Aubigné and Postel hip score to improve from 5 points to 14 points [9]. Lim et al. performed THA in 13 patients with confirmed diagnosis of MED and reported the Haris hip score improvement from 40.6 to 93.8 points [7]. However, amongst the 13 patients, only 3 underwent staged bilateral THA and the average followup was 4.8 years. In their study, Vanlommel et al. reported the outcomes of hybrid THA in seven MED patients with mean followup of 8.4 years [8]. They noted improvement in Charnley, Merle d'Aubigné and Postel hip scores following THA. However, only four of the seven underwent staged bilateral THA. Furthermore, it must be noted that two patients in their cohort had undergone either bilateral pelvic or femoral osteotomies prior to the THA. Ramaswamy et al. performed 16 THA (combination of uncemented and hybrid THA) in 9 patients with mean follow up of 15.9 years [6.] Overall, HO was noted in 11 hips and 10 hips required revision at an average follow up of 12.5 years. The mean Merle d'Aubigné and Postel hip scores improved from a mean of 6.4 points to 10.6 at latest follow up. The findings of the current study in terms of patient demographics, symptom characteristics, improvement in functional limitations following arthroplasty are in close agreement with the results of the published studies (Table 1, 2 and 3).

It has been demonstrated that in general patients with musculoskeletal dysplasia undergoing arthroplasty are at an increased risk of surgical site infection (SSI) [16]. Furthermore, complications such as SSI and HO have been reported following THA in patients with MED [6, 9, 11, 16]. Similar findings were noted in our cohort with the THA wounds healing over a longer duration in comparison to the TKA wounds. One patient (case 2) required wound debridement and subsequently developed HO (Figure 3). In contrast, all the TKA wounds in the three MED patients healed uneventfully. Currently, there is no

definitive explanation for the increased incidence of SSI or HO in this patient population. However, a combination of the prominent greater trochanter and altered anatomical features of the proximal femur and soft tissues around the hip region may play a role.

The current study has certain limitations in that it represents a small cohort of patients from a single specialist orthopaedic hospital. Image or sensor based navigation technology was not used during the procedures. However, conventional instrumentation and jigs which are routinely used in many hospitals were used in our patients with careful preoperative planning and implemented meticulously during surgery. The robust postoperative outcomes validate this approach. The healthcare model of our institution with extended inpatient stay which permitted staged procedures and specialist multidisciplinary input to optimise patient outcomes may not be widely applicable. Nonetheless, this study has demonstrated some of the salient principles which can be adopted to ensure good long term outcomes with bilateral lower limb arthroplasty in patients with MED.

5. Conclusions

Bilateral lower limb arthroplasty of the hip and knee is a good treatment option to restore function and mobility in MED patients with advanced degenerative arthritis. Careful preoperative planning, meticulous surgical techniques, patient focussed postoperative rehabilitation with multidisciplinary team approach are vital to ensure good functional outcomes and implant survivorship.

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