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The Modified West China Hospital Radiographic Classification for Fibrous Dysplasia in Femur: A Retrospective Analysis of 238 Patients

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Research article

Keywords: Fibrous dysplasia, Radiograph, Classification, Treatment

Posted Date: November 23rd, 2020

DOI: https://doi.org/10.21203/rs.3.rs-104794/v1

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Abstract

Background: To investigate the reliability and clinical outcome of a newly developed classification system for patients, who had fibrous dysplasia (FD) in the femur**.**

Methods: A total of 238 patients with FD in the femur were included in this retrospective study. All affected femurs were measured and treated based on our classification. The intraobserver and interobserver reproducibility were assessed using the Cohen kappa statistic**.** The clinical outcome was evaluated using the criteria of Guille.

Results: At a median follow-up of 60 months, 238 patients were categorized into the following five types: type I to V. The interobserver and intraobserver kappa scores were excellent. For clinical outcomes, there was no significant difference in the postoperative Guille score between type I (mean 9.01 ± 1.22), II (mean 8.40 ± 1.38), and V (mean 8.47 ± 1.69). Type III and IV had significantly lower postoperative Guille scores than type I, II, and V. Moreover, type III had a significantly higher Guille score (mean 7.81 ± 0.96) than type IV (mean 6.57 ± 2.09).

Conclusion: Our classification is reproducible and provides a one-to-one correspondence between diagnosis and treatment. Therefore, we recommend this classification for the diagnosis and treatment of the FD in the femur.

Introduction

Fibrous dysplasia (FD) is a common skeletal disorder (monostotic or polyostotic) that results in pathological fractures, deformity, limping, and pain. FD is a benign intramedullary fibro-osseous lesion, which was first identified by Lichtenstein in 1938[1]. The mutation of the GNAS-gene decreases the GTPase activity of the stimulatory G-protein, which increased the intracellular levels of cyclic adenosine monophosphate (cAMP) and interleukin-6 (IL-6) secretion[2]. The increased intracellular cAMP content and increased IL-6 secretion result in the increased numbers of osteoclasts and bone resorption. Primitive bone has failed to transform into the mature lamellar bone and realign in response to mechanical stress[3]. The lower extremity is frequently affected by deformity, fracture, leg length discrepancy (LLD), and limping[4–7]. Given the numerous surgical options and complicated deformities in the femur, it can be difficult for orthopedic surgeons to evaluate the severity of deformities and select an appropriate treatment strategy. Ippolito et al[8] have developed a classification system that characterizes femoral deformities of FD in six patterns, which only serve to guide the prediction of progression.

The surgical strategy is aimed at bone pain relief[9], restoring normal femoral alignment[5], gait normalization[6], LLD[5], and preventing pathological fracture[4, 10] for FD patients. Based on our early experience in assessing deformities and pairing treatment strategies[6, 11, 12], we modified our previous classification into five types following corresponding treatment options, which were as follows: including proximal femur bone loss, neck-shaft angle, femoral shaft deformity, genu valgum, and arthritis of hip. This study aimed to investigate the interobserver reliability and intraobserver reliability of our system and to evaluate the outcomes of surgical procedures based on our classification system that pair surgical strategy.

Methods

Development of our classification system

For radiographic analysis, the following features were determined in the lower extremity radiographs and axial computed tomography (CT) scans complex deformities based on standing limb alignment[13]: focal thinning of cortical bone and involvement of the calcar in the proximal femur, which measured on axial CT[14]; coxa vara, in which the neck-shaft angle was ≤ 120°; metaphyseal and femoral shaft deformity, in which varus or valgus malalignment was detected; genu valgum, in which the mechanical femorotibial angle was ≥ 10°[15]; hip arthritis, in which was detected according to the Kellgren Lawrence grading system[16].

Radiographic Management

Type I lesion was defined as none of the five features mentioned above. Type II lesion was defined as extensive bone loss of proximal femur (focal thinning of cortical bone and/or involvement of the calcar). Type III deformity included coxa vara and femoral shaft deformity, either alone or in combination. Additionally, hip internal or external rotation was measuring. Type IV deformity was characterized by severe genu valgum. And, type IV was to be alone or in combination with type III. Type V deformity was defined as any type associated with severe arthritis of the hip. For polyostotic FD patients involving the bilateral lower limbs, we record the severer side. To eliminate LLD, preoperative planning set the intact side (monostotic cases) or relatively short side (polyostotic cases) as standard. The opening- and closing-wedge osteotomy were provided for preventing LLD intraoperatively.

We retrospectively analyzed FD patients who were treated between January 2009 and January 2019 in our institution. Radiological diagnosis was made according to the features, including a grayish “ground-glass” appearance, endosteal scalloping, shepherd’s crook deformity, and intramedullary expansible lesion with a smooth sclerotic margin[13]. Biopsy with histological evaluation was required in questionable cases and patients with a high suspicion of malignancy. All type II-V cases were histologically proven FD postoperatively. The study was approved by the Institutional Ethics Committee of West China Hospital (Chengdu, China), and the study protocol adhered to the guidelines stipulated in the World Medical Association Declaration of Helsinki.

Perioperative Management

For correspondence treatment of each type, type I was monitoring every 6 months[10]. And the indication for surgical treatment in type II-V patients included the following: mechanical/ weight-bearing bone pain, hip and/or knee stiff, walking with a limp, fracture, and severe LLD[10]. Therefore, type II was treated with internal fixation (IF) following simple curettage[4]. Type III was treated with the IF following single or multiple level valgus osteotomies and simple curettage[5, 6]. Type IV was treated with a high tibial osteotomy (HTO)[17] or distal femoral osteotomy (DFO)[18], following femoral malalignment correction. Type V was treated with total hip arthroplasty (THA) and lesion curettage[9], if necessary single-level valgus osteotomy was performed to fit femur stem[12] (Table 1)

Table 1
Radiographic Classification System of Fibrous Dysplasia in Femur

Type	Extensive bone loss of proximal femur	Coxa vara	Femoral shaft deformity	Genu valgum	Hip arthritis	Intervention
Type I	-	-	-	-	-	Monitoring
Type II	+	-	-	-	-	Curettage + IF
Type III	+/-	+/-	+/-	-	-	Curettage + Single/Multiple-level osteotomy + IF
Type IV	+/-	+/-	+/-	+	-	Curettage + Single/Multiple-level osteotomy + IF + HTO or DFO
Type V	+/-	+/-	+/-	+/-	+	THA +/- Single-level osteotomy
IF, internal fixation;						
THA, total hip arthroplasty						

Type I patients followed up with a semi-annual assessment. Early weight-bearing protocols varied in type II-IV. Type II patients were allowed full weight-bearing immediately after surgery. Type III and IV patients were allowed toe-touch weight-bearing within 6 weeks, and partial weight-bearing at 6–12 weeks postoperatively. Then progressive weight-bearing was permitted thereafter[19]. For type V patients who underwent valgus osteotomy, the weight-bearing protocol was the same as that of type III and IV. Without valgus osteotomy, the protocol of type V patients was the same as that for type II patients[12].

Agreement Analysis

In the specialist group, the mean interobserver and intraobserver kappa scores were 0.85 (range 0.80–0.89) and 0.87 (0.79–0.92), respectively. In the resident group, the mean interobserver and intraobserver kappa scores were 0.80 (range 0.77–0.81) and 0.83 (0.80–0.84), respectively. In total, the mean interobserver and intraobserver kappa scores were 0.85 (range 0.77–0.89) and 0.85 (0.79–0.92), respectively. The highest percentage of erroneous classification was observed in types I and II.

Results

Classification of our population

A total of 238 patients with FD in the femur were enrolled in our institution. Of the included 238 patients, 168 had monostotic FD and 70 had polyostotic FD. All femurs could be categorized by our radiographic classification. The classification including five types of FD was noted (Table 2).

Table 2
Patient Characteristics*

	No. of Patients (%)
Age (years)	median 29 (range 7.1–73)
Gender	
Female	141(59.2)
Male	97(40.8)
Category	`
Monostotic	168(70.6)
Polyostotic	70(29.4)
Prevalence of different types	
Type I	82(34.5)
Type II	72(30.3)
Type III	48(20.2)
Type IV	21(8.8)
Type V	15(6.3)
Follow-up (months)	
Total	median 60 (range 6-120)
Type I	median 63 (range 6-120)
Type II	median 48 (range 12–120)
Type III	median 72 (range 60–120)
Type IV	median 60 (range 24–120)
Type V	median 55(range 23–65)
*N = 238 patients (250 affected femurs)	

Clinical Outcome

At a median follow-up of 60 months (range 6-120 months), all patients were evaluated using the criteria of Guille et al[6, 21]. With regards to the variations of functional outcomes before and after surgery, there was a significant increase in the postoperative Guille score in types II-V, compared to the preoperative values. In type I, there was no significant difference between the initial and latest assessments. As for the postoperative Guille score, there was no significant difference among type I (mean 9.01 ± 1.22), II (mean 8.40 ± 1.38), and V (mean 8.47 ± 1.69) cases. Type III and IV cases were significantly lower postoperatively Guille score than type I, II, and V. Moreover, type III cases had significantly higher Guille score (mean 7.81 ± 0.96) than type IV cases (mean 6.57 ± 2.09) (Table 3).

Table 3
Preoperative/postoperative clinical scores evaluated by the modified criteria of Guille

Categories	Type II(n = 72)				Type III(n = 48)				Type IV(n = 21)				Type V(n = 1	
	U* (n)	A* (n)	S* (n)	Mean score	U* (n)	A* (n)	S* (n)	Mean score	U* (n)	A* (n)	S* (n)	Mean score	U* (n)	A* (n)
Pain	49/1	23/14	0/57	0.32 ± 0.47/1.78 ± 0.45	46/2	2/12	0/34	0.13 ± 0.33/1.67 ± 0.56	16/2	5/7	0/12	0.24 ± 0.44/1.48 ± 0.68	15/2	0/1
Hip motion	22/2	38/24	12/46	0.86 ± 0.68/1.61 ± 0.55	39/0	9/21	0/27	0.19 ± 0.39/1.56 ± 0.50	10/3	10/8	1/10	0.67 ± 0.66/1.33 ± 0.73	15/0	0/4
Limping	43/0	10/19	19/53	0.67 ± 0.87/1.74 ± 0.44	41/1	3/22	4/25	0.23 ± 0.59/1.50 ± 0.55	10/2	5/11	6/8	0.81 ± 0.87/1.29 ± 0.64	13/1	2/4
Activities of daily living	51/0	6/21	15/51	0.50 ± 0.82/1.71 ± 0.46	45/1	2/18	1/29	0.83 ± 0.35/1.58 ± 0.54	9/0	9/12	3/9	0.71 ± 0.72/1.43 ± 0.51	12/0	3/2
Social activities	56/1	9/29	7/42	0.32 ± 0.65/1.57 ± 0.53	47/2	1/20	0/26	0.21 ± 1.44/1.50 ± 0.58	12/3	9/14	0/4	0.43 ± 0.51/1.05 ± 0.59	15/1	0/4
Total				2.67 ± 2.44/8.40 ± 1.38				0.65 ± 0.98/7.81 ± 0.96				2.86 ± 1.98/6.57 ± 2.09		

*U = unsatisfactory, A = average, and S = satisfactory. Clinical outcomes were scored as 0 (unsatisfactory), 1 (average), or 2(satisfactory). For a potential max points, >9 points were defined as excellent, 7 or 8 points as good, 5 or 6 points as fair, and <5 points as poor.

For complications, two type III patients still complained of pain, one type III patients had mild-to-moderate Trendelenburg gait. One type IV patients had a mild Trendelenburg gait. And two type V patients still had mild limping.

Discussion

This study provides a framework for the systematic evaluation and management of the FD-induced-deformity. The spectrum of femoral deformities in full-length was classified into five categories. Furthermore, their corresponding management was recommended. Currently, there have been only two prior systematic categories[8, 11], which classify a variety of proximal femoral deformities in FD. However, both classifications were failed to guide the most appropriate strategy for treatment. In Ippolito et al's[8] study, three orthopedic surgeons and one pathologist evaluated FD femurs on two occasions with an interval of 6 weeks. The intraobserver (0.855) and interobserver (range, 0.833–0.871) agreement were both excellent. The highest percentage of mistakes was made when distinguishing mild shepherd's crook deformity from severe shepherd's crook deformity. In our primary classification system[11], two senior orthopedic surgeons evaluated the cases for two rounds with a 6-week interval. The intraobserver and interobserver agreements were both excellent. In this study, the mean interobserver and intraobserver kappa scores were 0.85 (range 0.77–0.89) and 0.85 (0.79–0.92), respectively. Although the agreement was excellent, the dispute was focused on the genu valgum between types III and IV. In some of the type IV lesions, genu valgum secondary to the restoration of the normal femoral alignment was remarkable after valgus osteotomy. Therefore, the surgeon must be prepared to manage the genu valgum in some type IV patients. Moreover, we recommend classifying these patients into type IV for correspondence surgical options (Fig. 1).

Type I lesions have no focal thinning of cortical bone and involvement of the calcar in the proximal femur, and we recommend conservative treatment with monitoring every 6 months (Fig. 2). Bone pain in FD should be discreetly assessed. Focal or weight-bearing pain may indicate an imminent or impending fracture[22]. Physiotherapy and pain medication can be administered, including opioids and non-steroidal anti-inflammatory drugs[10]. Besides, intravenous bisphosphonate is proposed for persistent, moderate to severe pain, even in children and adolescents[23, 24]. Moreover, denosumab, targeting RANKL that is expressed by osteogenic cells, maybe a potential treatment for bone pain caused by FD[25]. During follow-up, no severe complications were detected in type I patients.

Type II lesions are characterized by focal thinning of cortical bone and/or involvement of the calcar, without other femoral deformities. Type II patients often have mechanical or weight-bearing bone pain, which is a signal of stress or an impending fracture[10, 26]. Therefore, curettage, bone graft, and internal IF are recommended (Fig. 3). The efficacy and complications of bone graft are still controversial[10, 27]. However, cortical allografts were still recommended for the final and slow internal replacement by the host bone, especially in monostotic FD patients[3, 5]. The intramedullary lesion should be adequately bridged by IF including dynamic hip screw (DHS), anatomical plate, or intramedullary nail[4, 28, 29]. In our study, only one type II patients had mild-to-moderate pain postoperatively.

Type III patients refer to the deformity of coxa vara and/or femoral shaft deformity associated with bone pain. For the single-level osteotomy site, the subtrochanteric region[5, 6, 30] (Fig. 4) and the dome of the deformity[6, 31, 32](Fig. 5) were recommended. However, the double-level osteotomy is strongly considered inadequate for correcting severe deformity[6, 31, 33]. After osteotomy, orderly curettage, massive impaction allograft, and IF are performed[4, 34, 35]. The choice of IF is still controversial. Previously, some authors suggested the longer DHS rather than intramedullary nailing, because of its ability in correcting varus and rotational deformities of the femoral neck and simplify procedure[5, 30, 32]. However, some studies report that intramedullary nails can provide good biomechanical support[6, 34]. In general, we recommend intramedullary nail for the following reasons. Firstly, it provides sufficient stability that prevents stress fracture and screw loosening or pullout, especially in polyostotic FD patients[5, 29]. Secondly, it accommodates multiple-level osteotomy[31]. To increase the initial stability, the transversal surface of the femur after osteotomy should be entirely matched for locking each other. Additionally, the intramedullary lesions with sclerotic rim have sufficient bone mass, which provides adequate stability for nail fixation. Therefore, the sclerotic bone should be discreetly preserved when curettage and reaming canal. Moreover, rotational deformities of the femur could be gradually corrected. In our study, two type III cases still complained of pain, remained pain, owing to mild hip joint degeneration. One type III patient had mild-to-moderate Trendelenburg gait. Preoperatively, the neck-shaft angle of this patient was only 75°. Over-tensioning of the gluteus medius was inevitable postoperatively after the correction of coxa vara[36].

Type IV are detected in patients with severe genu valgum. When combined with type III deformity, a two-stage treatment was recommended. Complex femoral deformities and lesion curettage were corrected firstly. After six months of rehabilitation, the second-stage procedure was performed for patients, who still complained of typical symptoms. We suggest HTO or DFO, for achieving a satisfactory appearance, correcting limb alignment, and relieving pain[18, 37]. However, for polyostotic FD patients, genu valgum is mainly caused by the proximal tibia and distal femur; thus, DFO and HTO are both recommended (Figs. 6 and 7). Type IV lesions are uncommon and the most challenging type and patients with this type of deformity have a lower Guille score, compared to other types of patients. In our study, only one type IV patient had mild Trendelenburg gait, because of the over-tensioning of the gluteus medius.

Type V lesions are found in FD patients with severe hip arthritis, combined with other types (II/III/IV). The deformities of the femoral shaft, coxa vara, and genu valgum, are significantly associated with the degeneration of the hip and knee. Additionally, polyostotic FD is found to be more prevalent in hip arthritis than monostotic FD[38, 39]. In our study, type V patients had significantly higher Guille scores than type III and IV, but no significant difference was found between patients with type V and those types I and II. Although a small fraction of FD patients with mild deformity may be classified into type V, the relatively high Guille score was preliminary evidence to guide treatment. Sierra et al.[9] firstly reported THA in patients with FD. When hip arthritis has been severe enough, total hip arthroplasty is suggested. Furthermore, a cemented stem was suggested to have a lower revision rate than an uncemented stem. However, our institution reported that the long uncemented stem showed reliable fixation at mid-term follow up[12], using Mimics V17.0 Software (Materialise Corp. Belgium), precise preoperative planning, and simulation of the osteotomy. Also, implantation of the prosthesis stem in a three-dimensional reconstructive model is essential. For the femoral component, we recommend fully coated stems, which engage in normal diaphyseal bone bypassing the lesion areas at least 2 femoral canal diameters to decrease the risk of postoperative fractures[9] (Figs. 8 and 9). The stem implantation is a crucial point. First, precision osteotomy was assisted by a patient-specific instrument, using a micropendulum saw. Second, the femoral cavity was temporarily fixed after thoroughly evacuation and bone graft. Third, the femur was reamed following addressing the greater trochanter. Two type V patients had mild limping, because of leg length discrepancy after THA.

There were several limitations to our study. Firstly, the classification was retrospective and confined to radiographic images. Secondly, the follow-up time was significantly different among the five groups. Thus, multicenter studies with a longer follow-up time are needed to make sufficient conclusions on our classification, especially for type V. Thirdly, surgical strategies and classifications are appropriate only for adult patients since most patients in our department were adults (median age 29 years, range 7.1–73 years). Finally, although many patients firstly visited the surgical department due to a pathologic fracture, we have treated some patients with conditions that may not be classified by our system.

Conclusions

We developed a new classification system for FD in the femur which was built on a review of the literature and clinical outcomes. Agreement analysis of the classification of our population showed that our classification system is reproducible, and clinically directed for standardizing the surgical treatment for these deformities. Moreover, follow-up evaluations showed pain relief and gait improvement in most patients. We believe that our classification system provides a one-to-one correspondence between diagnosis and treatment. Therefore, we recommend this classification for the diagnosis and treatment of the FD in the femur.

Abbreviations

FD: fibrous dysplasia; cAMP: cyclic adenosine monophosphate; IL-6: interleukin-6; LLD: leg length discrepancy; CT: computed tomography; IF: internal fixation; HTO: high tibial osteotomy; DFO: distal femoral osteotomy; THA: total hip arthroplasty; DHS: dynamic hip screw.

Declarations

Acknowledgements

We are thankful for the support of the nurse team from the Department of Orthopedics, West China Hospital. We are thankful for the patients enrolled in this study.

And we thank Xianliang Zhang M.D (Department of pathology, West China Hospital) for pathology evaluation.

Authors’ contributions

TYW, LM, and CQT were involved with the concept and design of this manuscript. YZ,TYW , WLZ and DH were involved with the acquisition of subjects and data. MXL, JW, YQZ, YL, and CQT were involved in the perioperative management. YTW, LM and CQT were involved in postsurgical evaluation of the patients. All authors contributed toward data analysis, drafting and critically revising the paper, gave final approval of the version to be published, and agree to be accountable for all aspects of the work. All authors read and approved the final manuscript.

Funding

This work was supported, in part, by the National Key Research and Development Program of China (No. 2016YFC1102003, Dr. Tu), and the Science and Technology Research Program of Sichuan Province (No. 2020YFS0036, Dr. Tu); the Chengdu Science and Technology Program Projects (No. 2017-CY02-00032-GX, Dr. Zhou) and the National Natural Science Foundation of China (No. 81801852, Dr. Zhou); the National Key Research and Development Program of China (No. 2017YFB0702604, Dr. Min).

Availability of data and materials

The datasets used and analyzed during the current study are available from the corresponding author on reasonable request.

Ethics approval and consent to participate

This study was approved and monitored by the Ethical Committee of West China Hospital, Sichuan University in China (No.2019342). All patients signed the informed consent.

Consent for publication

Written informed consent was obtained from all patients for publication of this study and any accompanying images.

Competing interests

The authors report no competing interest in this work.

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Figures

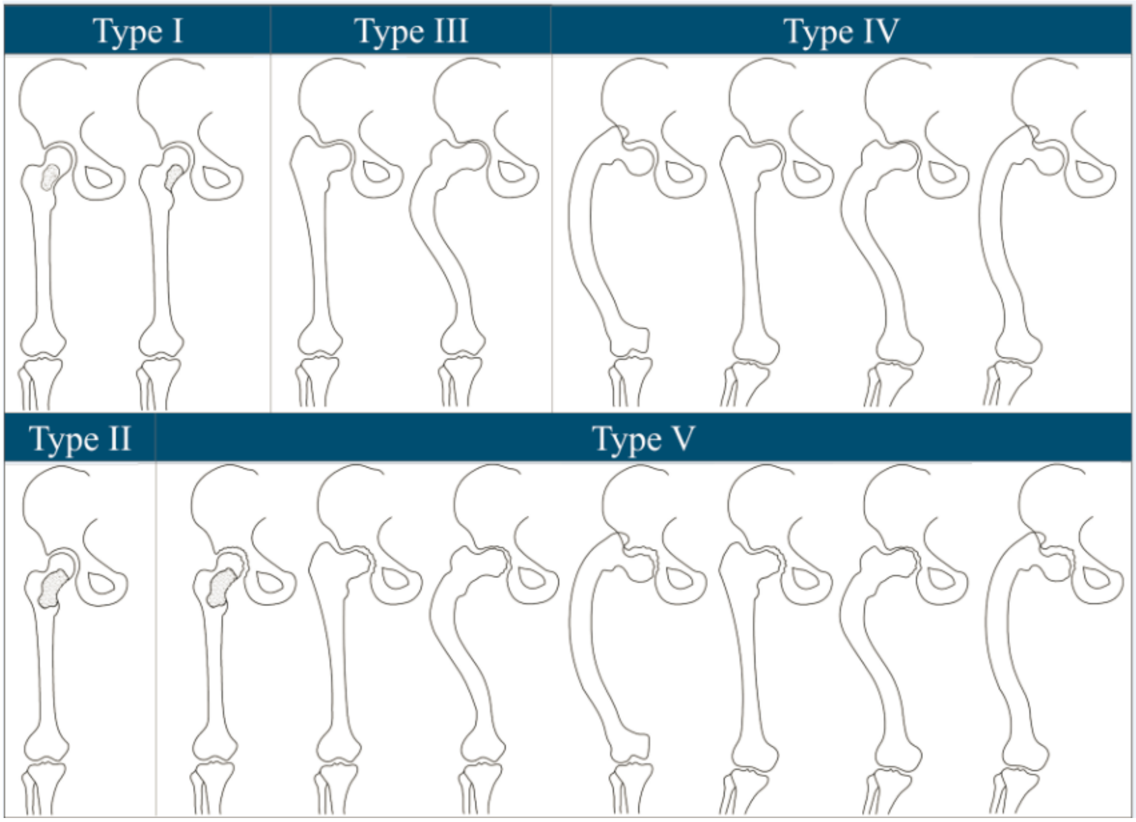
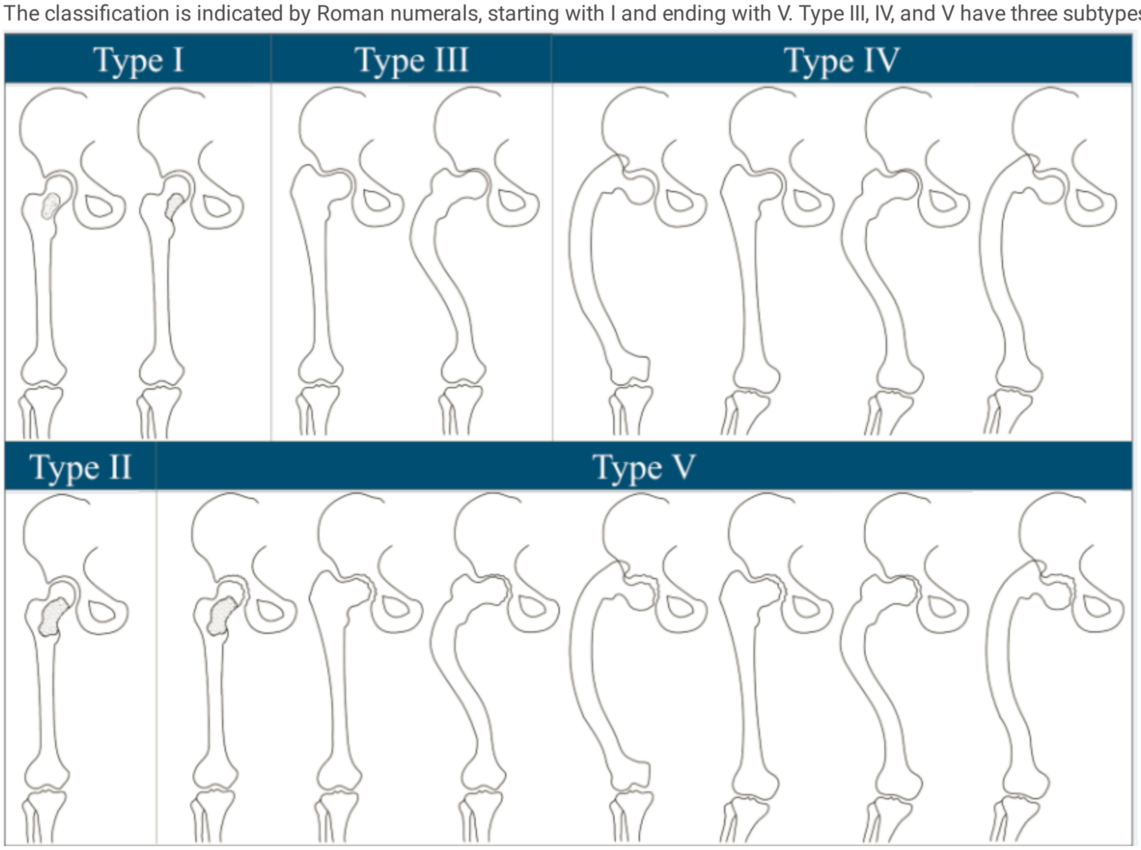


Figure 1

The classification is indicated by Roman numerals, starting with I and ending with V. Type III, IV, and V have three subtypes (A, B, and C).



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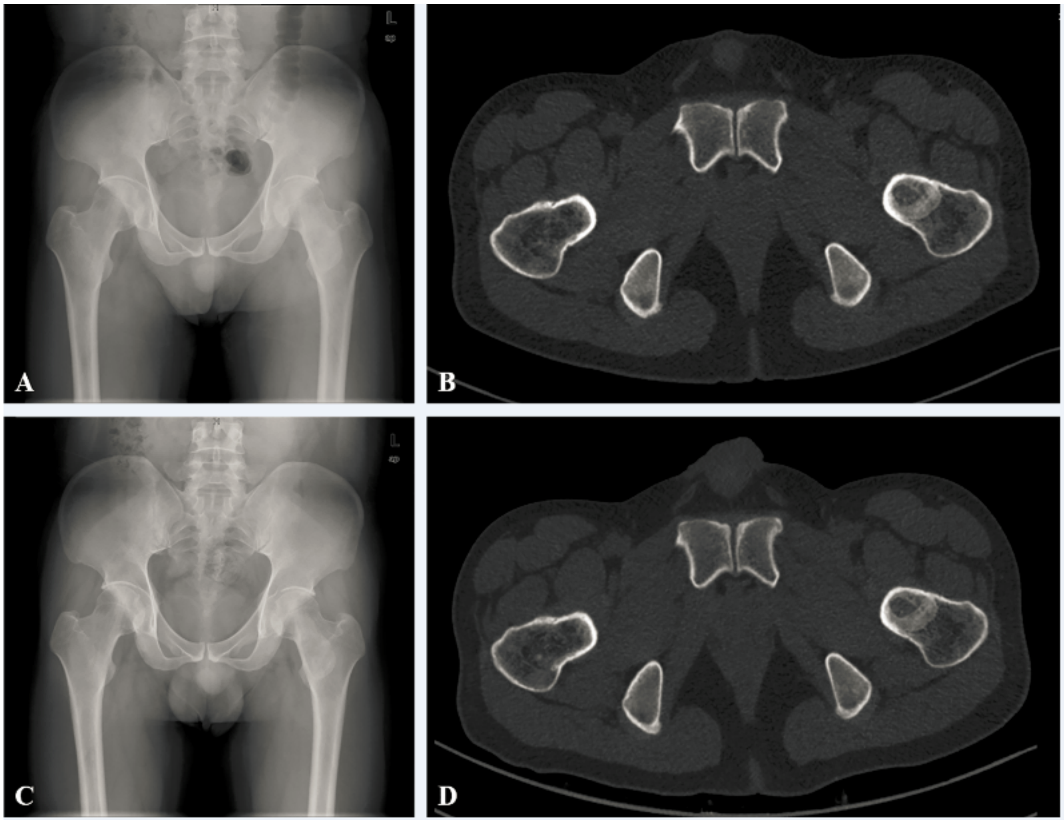
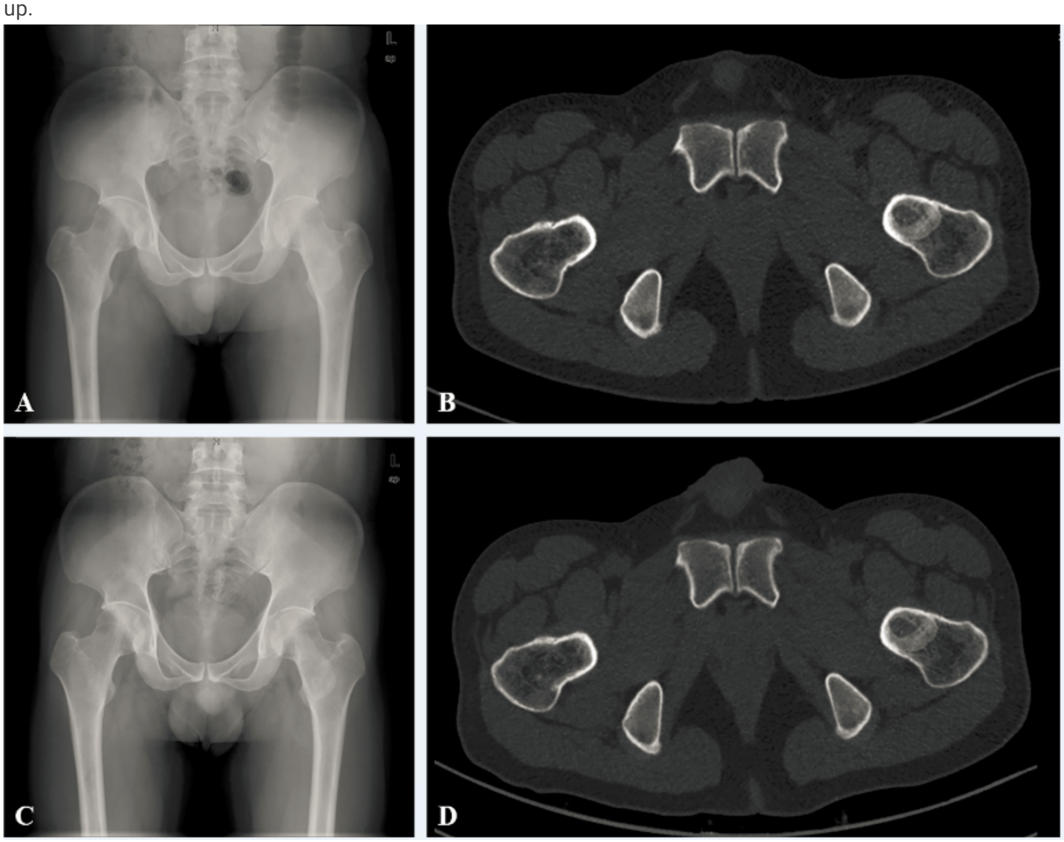


Figure 2

A and B Type I lesion was present in the left femoral neck with intact cortical bone and calcar. Fig. 2-C and D Type I lesion was latant at the 24-month follow-up.



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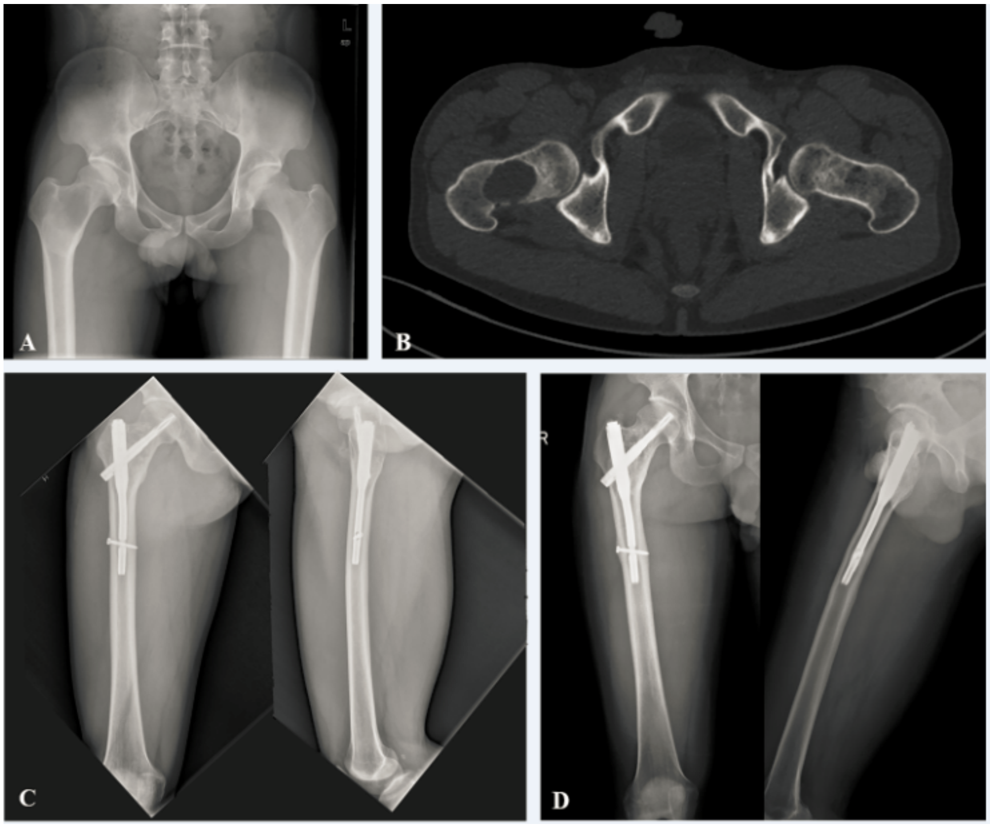
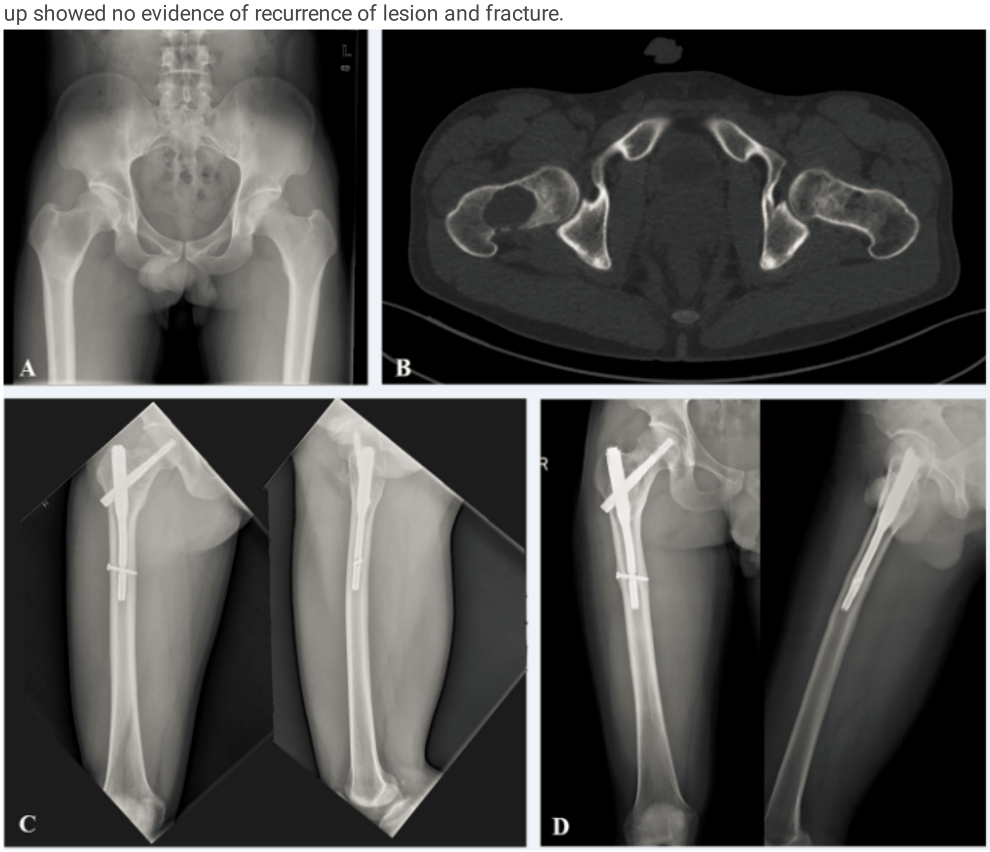


Figure 3

A Type II lesion. Fig 3-B The extensive bone loss of proximal femur (focal thinning of cortical bone). Fig 3-C 6 months after surgery. Fig 3-D 68-month follow-up showed no evidence of recurrence of lesion and fracture.



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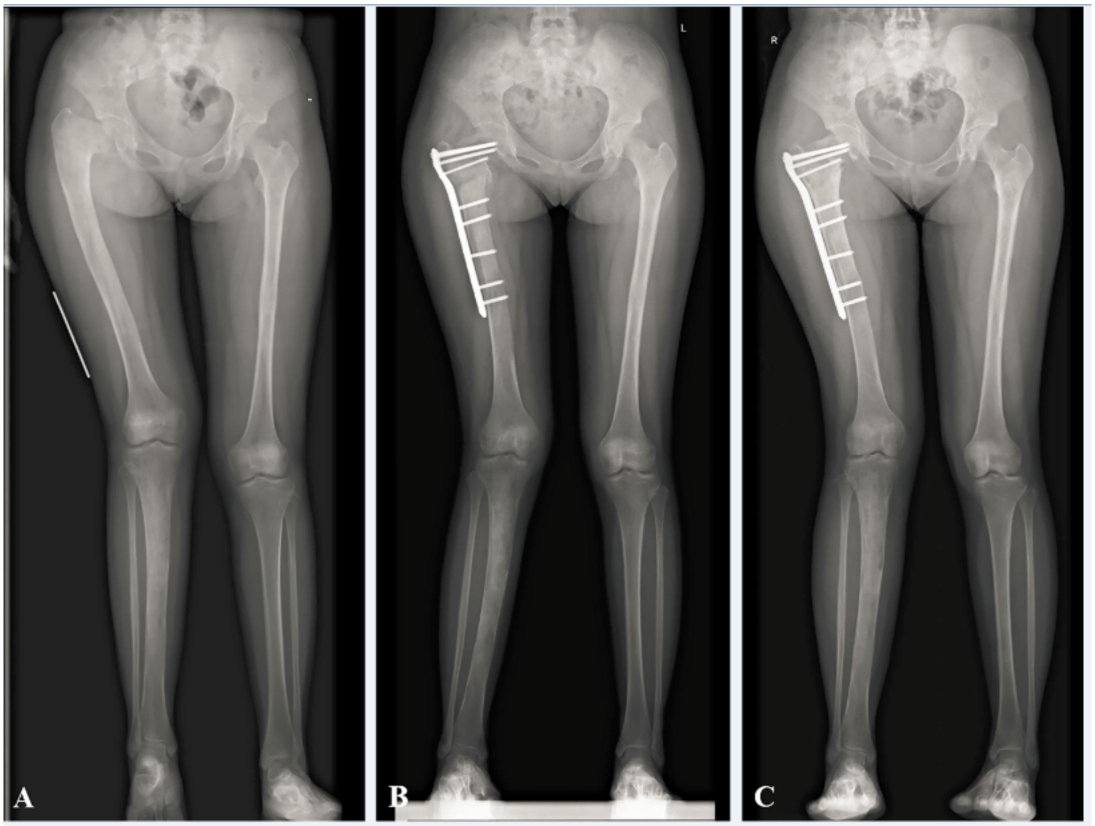
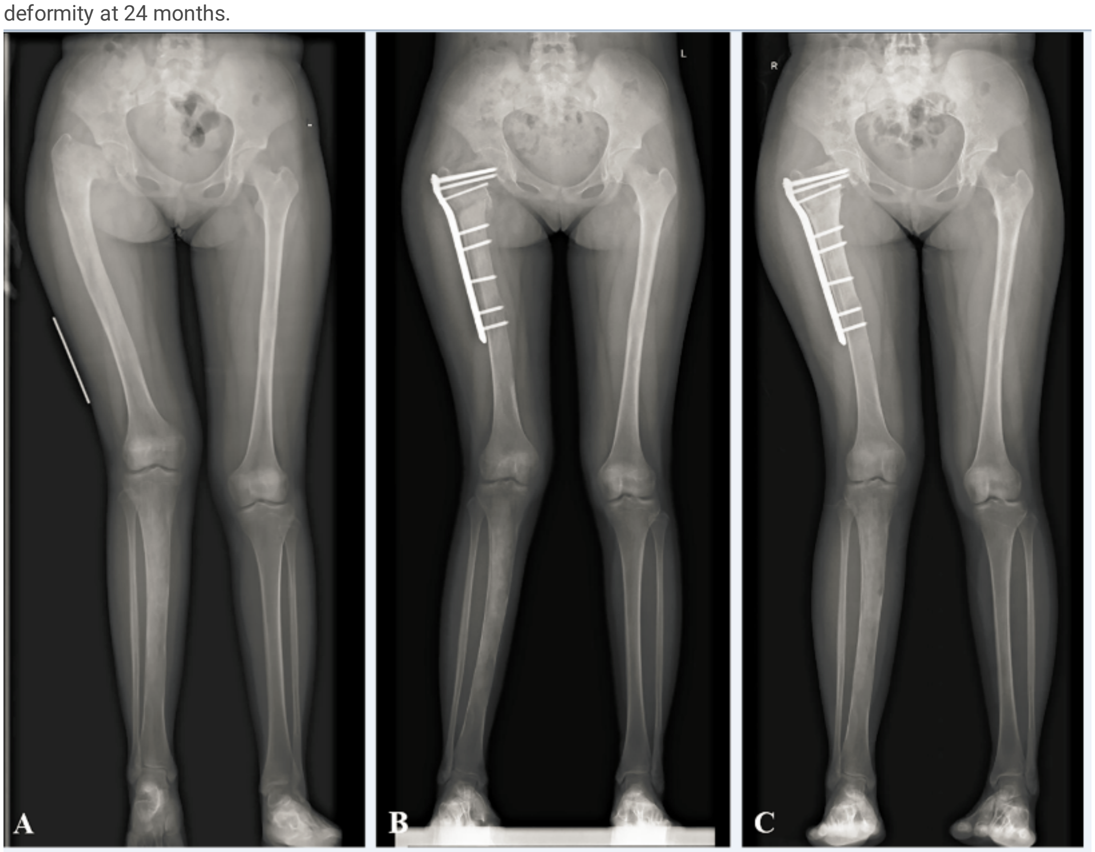


Figure 4

A Type III lesion with coxa vara. Fig. 4-B 7 month after surgery. Fig. 4-C The radiograph showed no evidence of recurrence of lesion and re-progress of deformity at 24 months.



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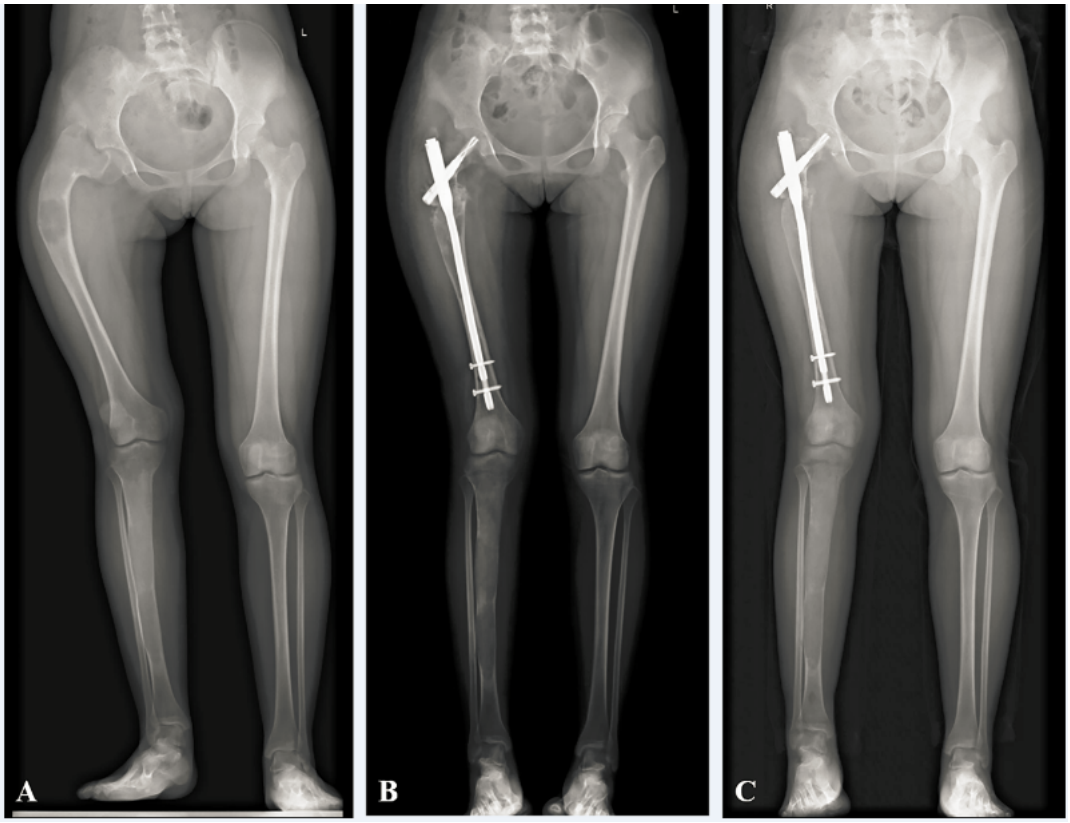
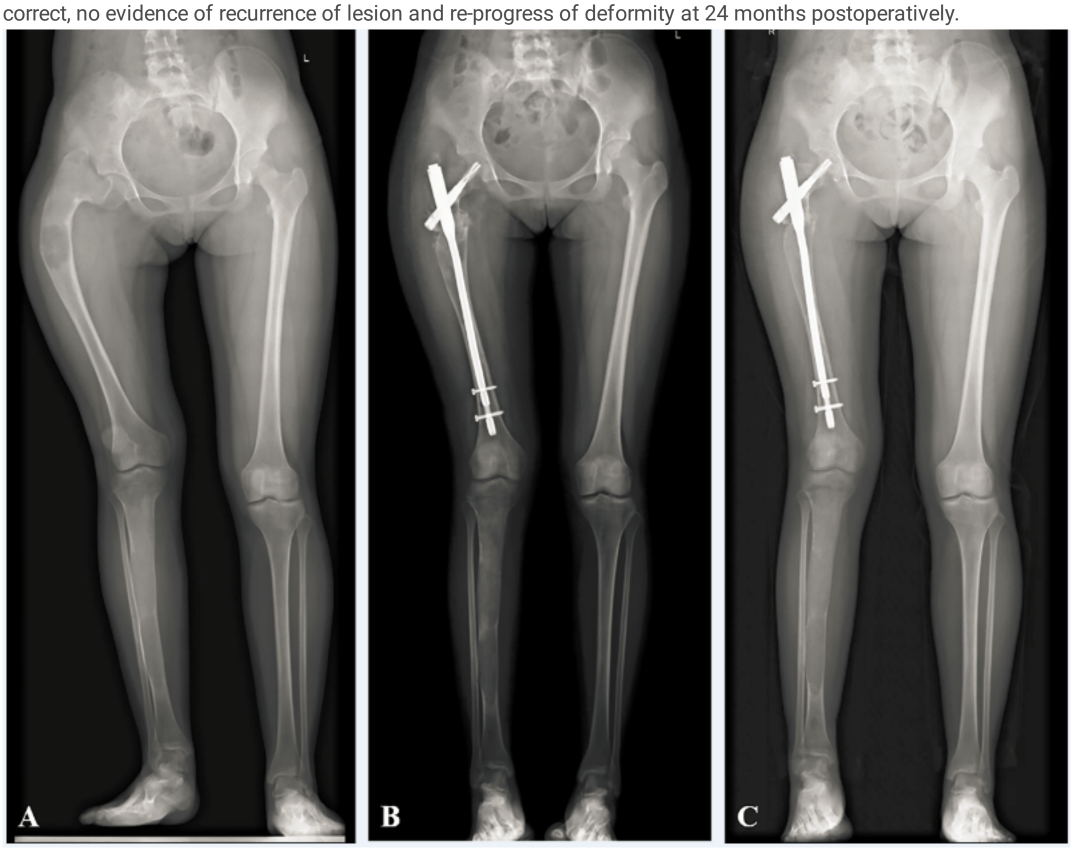


Figure 5

A Type III lesion. Fig. 5-B The radiograph at 2 months postoperatively. Fig. 5-C The radiograph showed good union of osteotomy site, alignment has been correct, no evidence of recurrence of lesion and re-progress of deformity at 24 months postoperatively.



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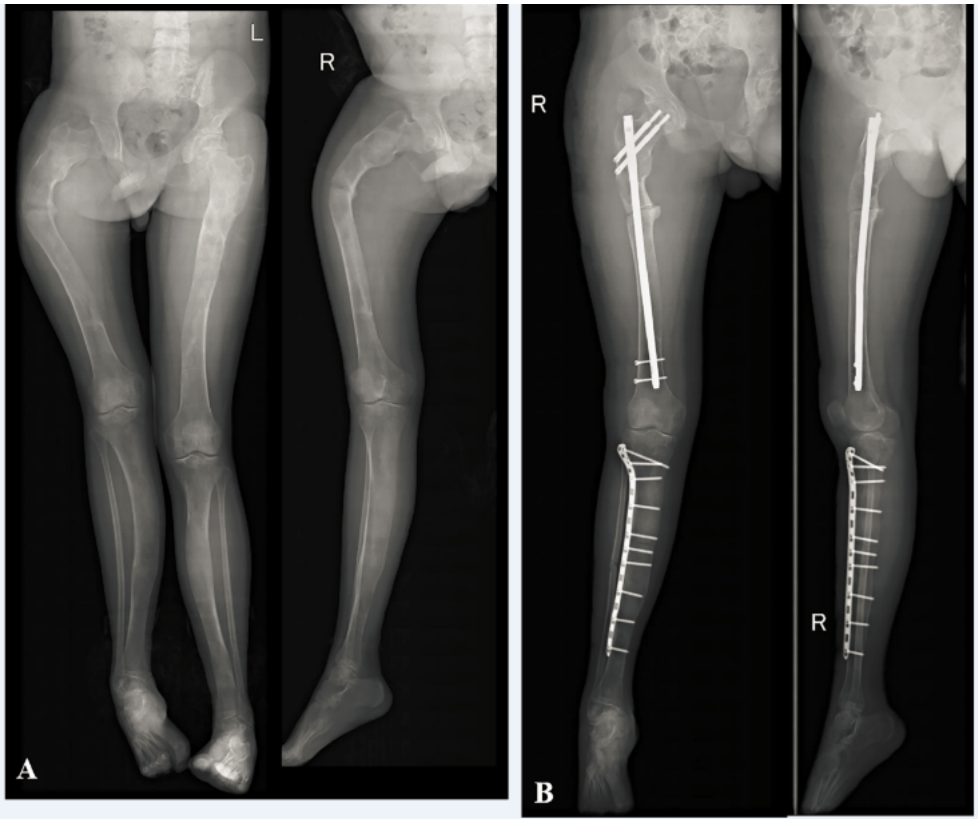
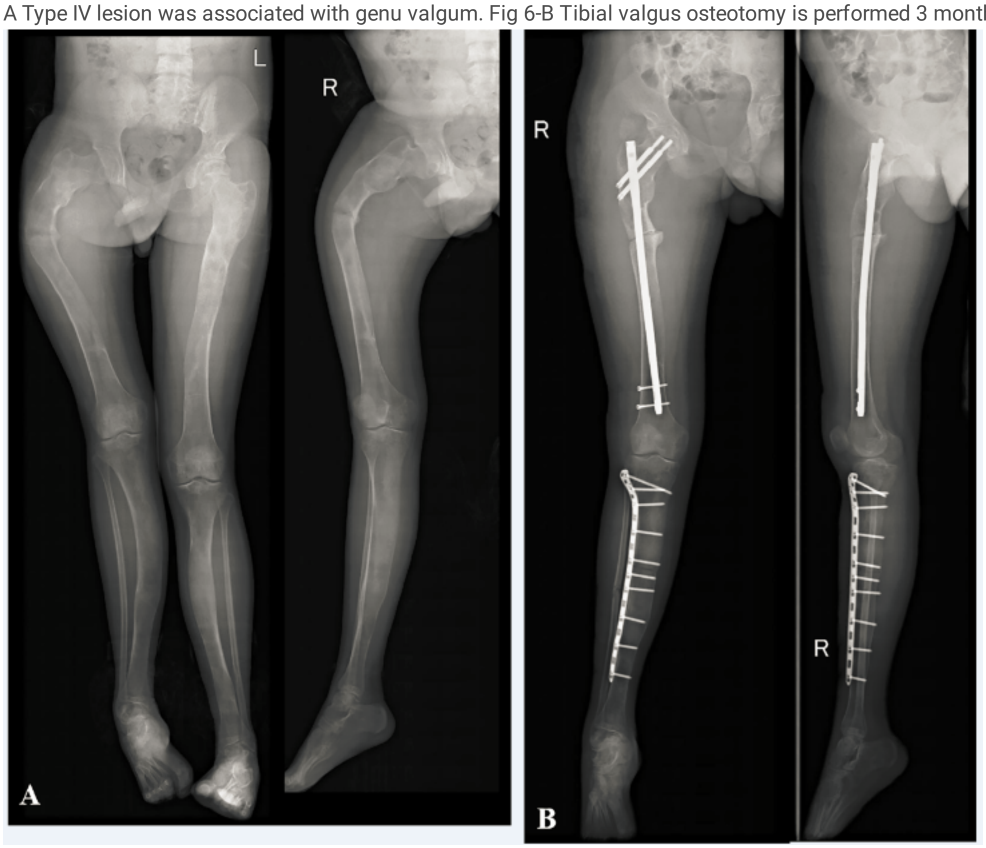


Figure 6

A Type IV lesion was associated with genu valgum. Fig 6-B Tibial valgus osteotomy is performed 3 months after the femoral valgus osteotomy.



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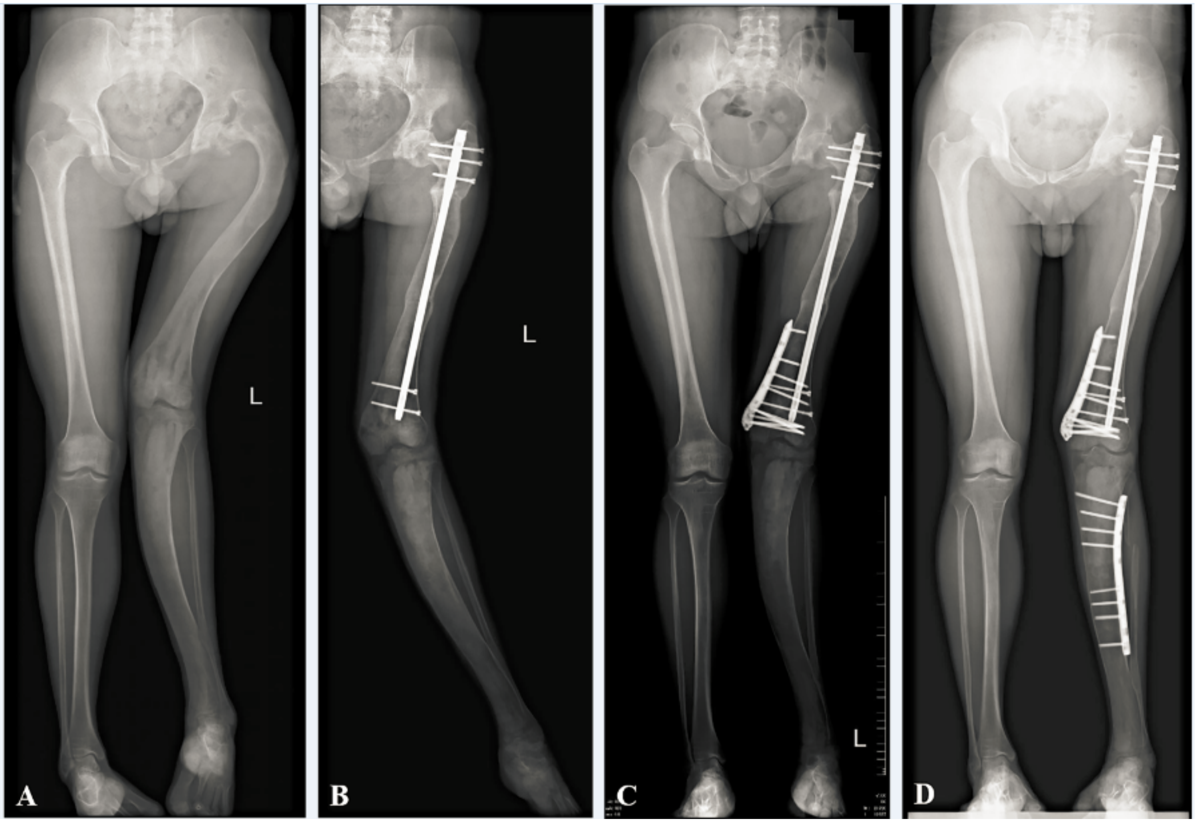
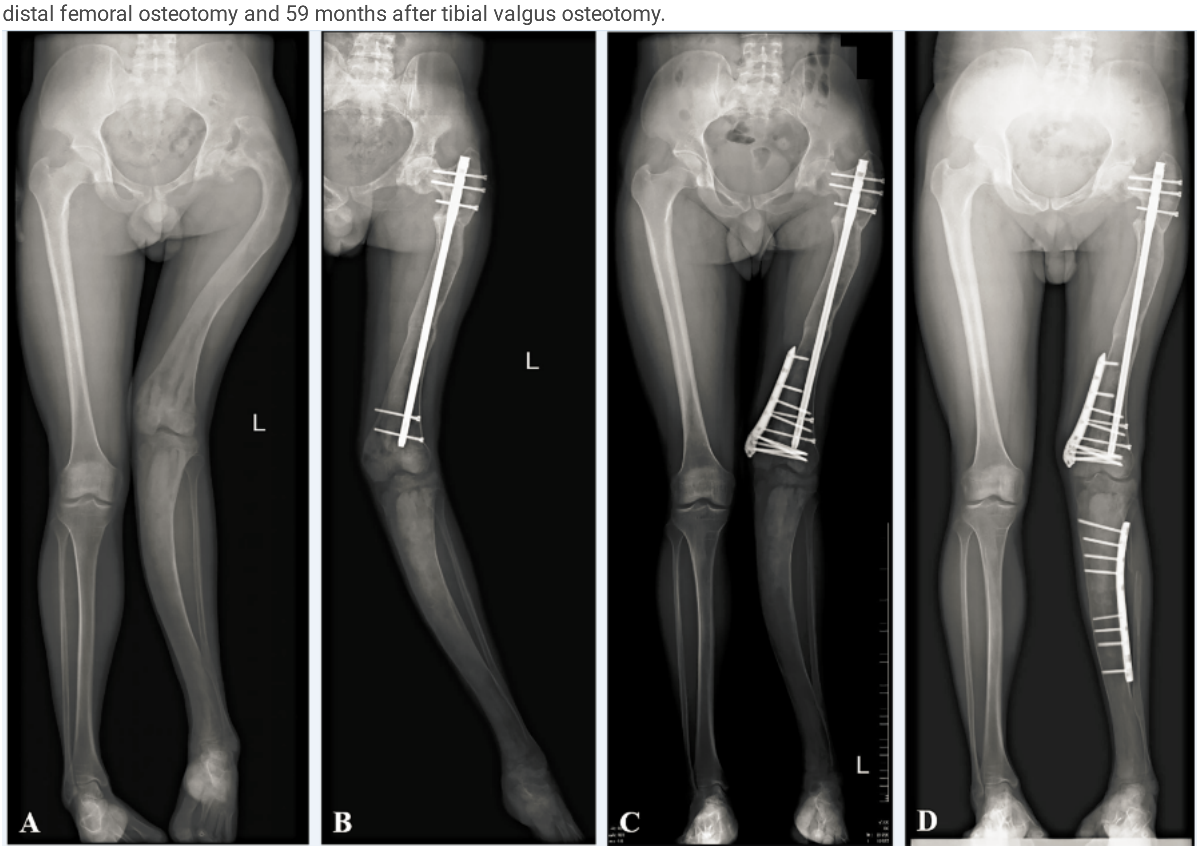


Figure 7

A Type IV lesion was associated with coxa vara, femoral shaft deformity, and genu valgum. Fig 7-B 9 months after the double-level osteotomy. Fig 7-C 21 months after the double-level osteotomy and 10 months after distal femoral osteotomy. Fig 7-D 80 months after the double-level osteotomy, 69 months after distal femoral osteotomy and 59 months after tibial valgus osteotomy.



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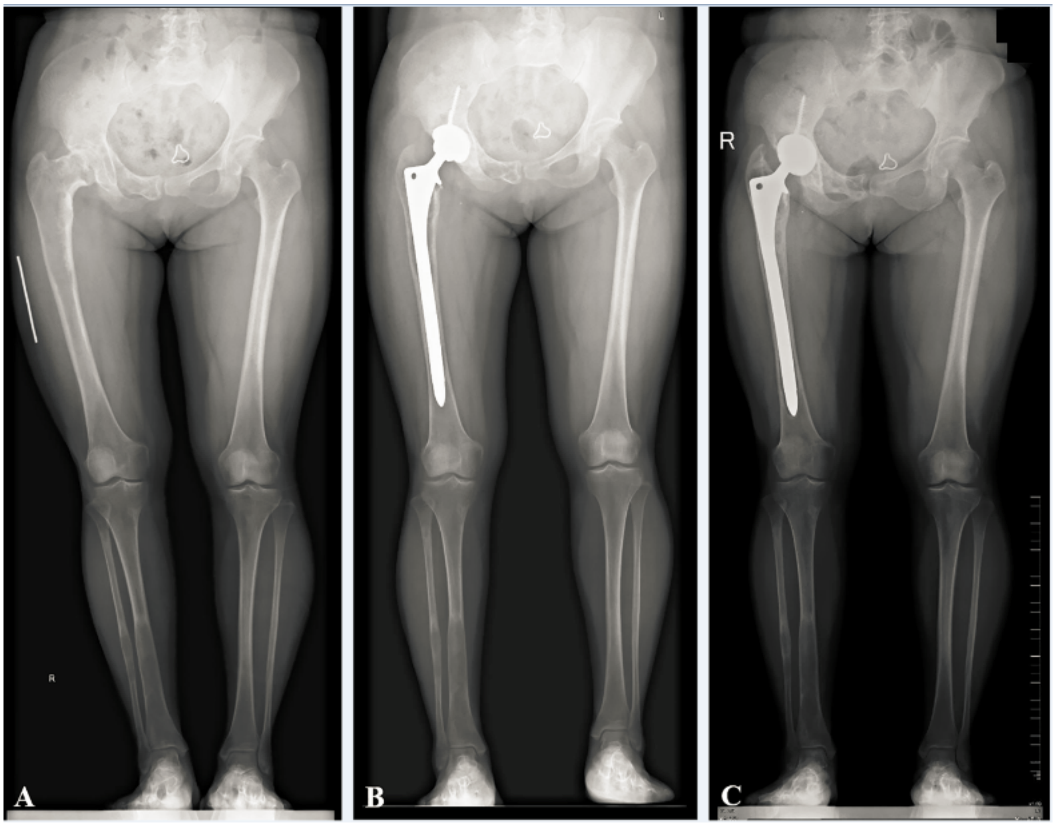
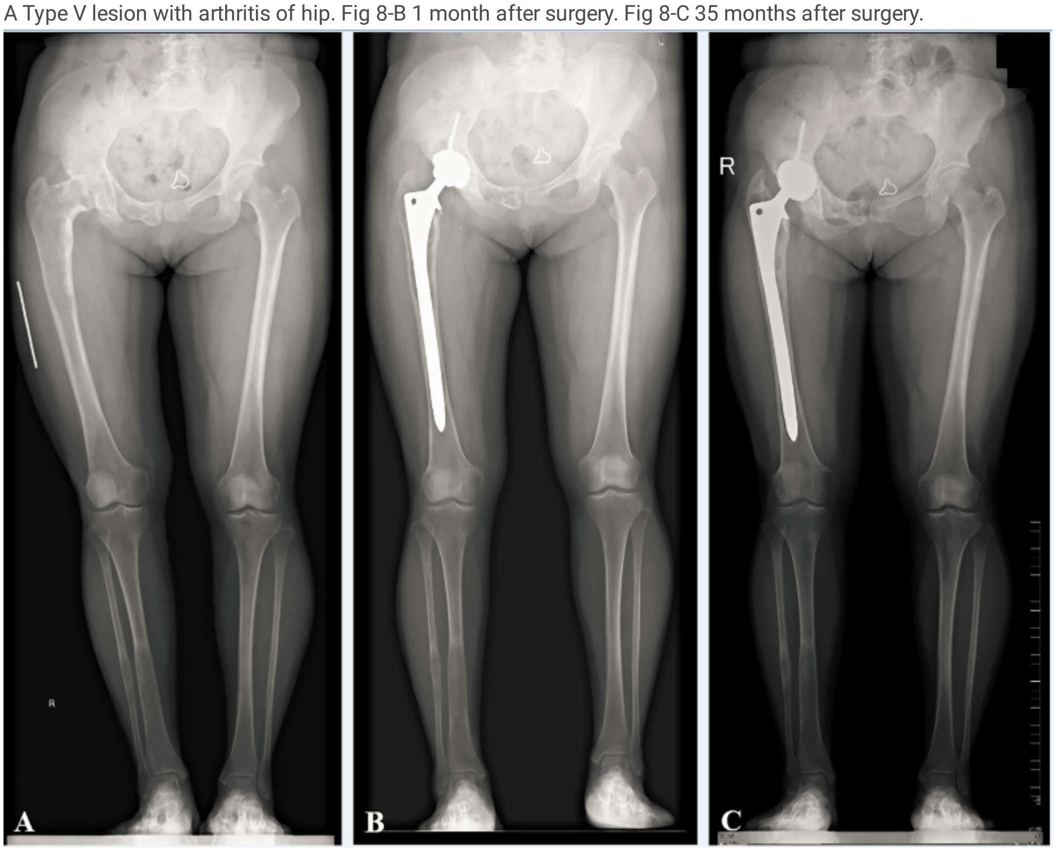


Figure 8

A Type V lesion with arthritis of hip. Fig 8-B 1 month after surgery. Fig 8-C 35 months after surgery.



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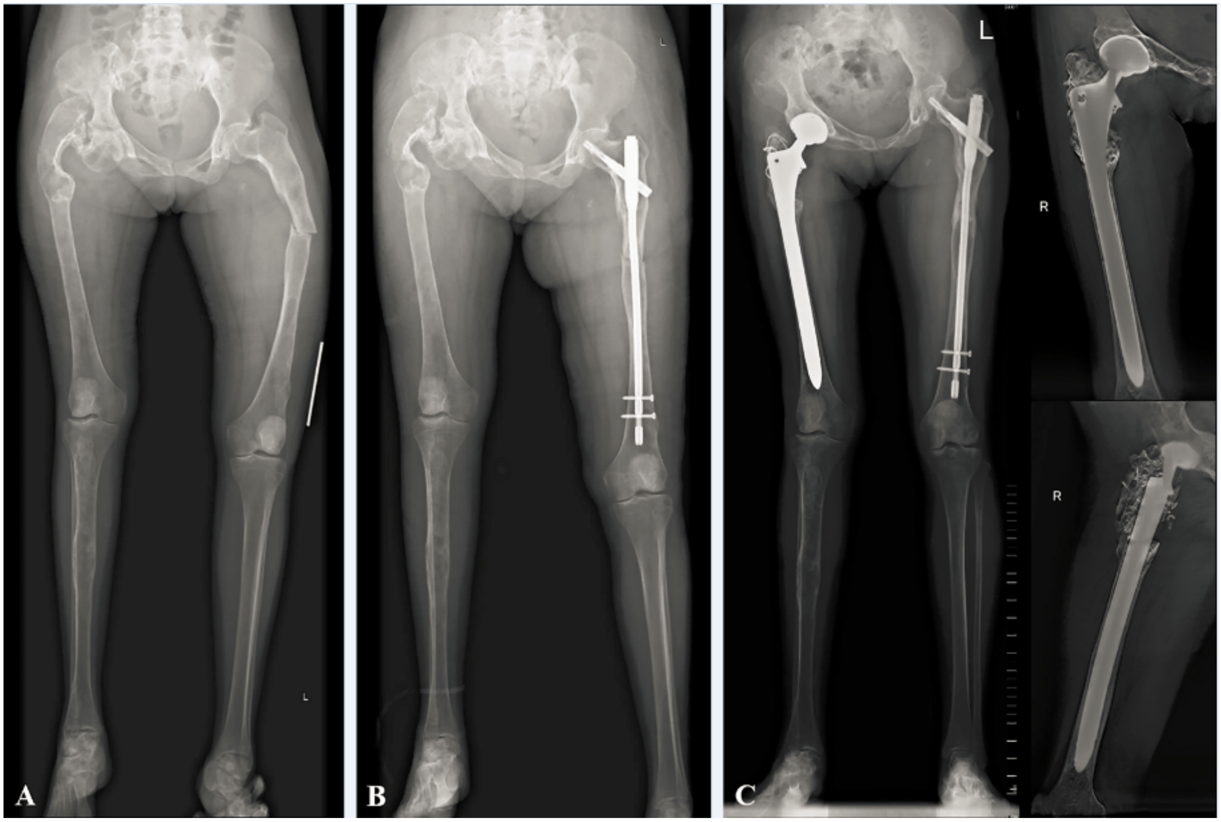
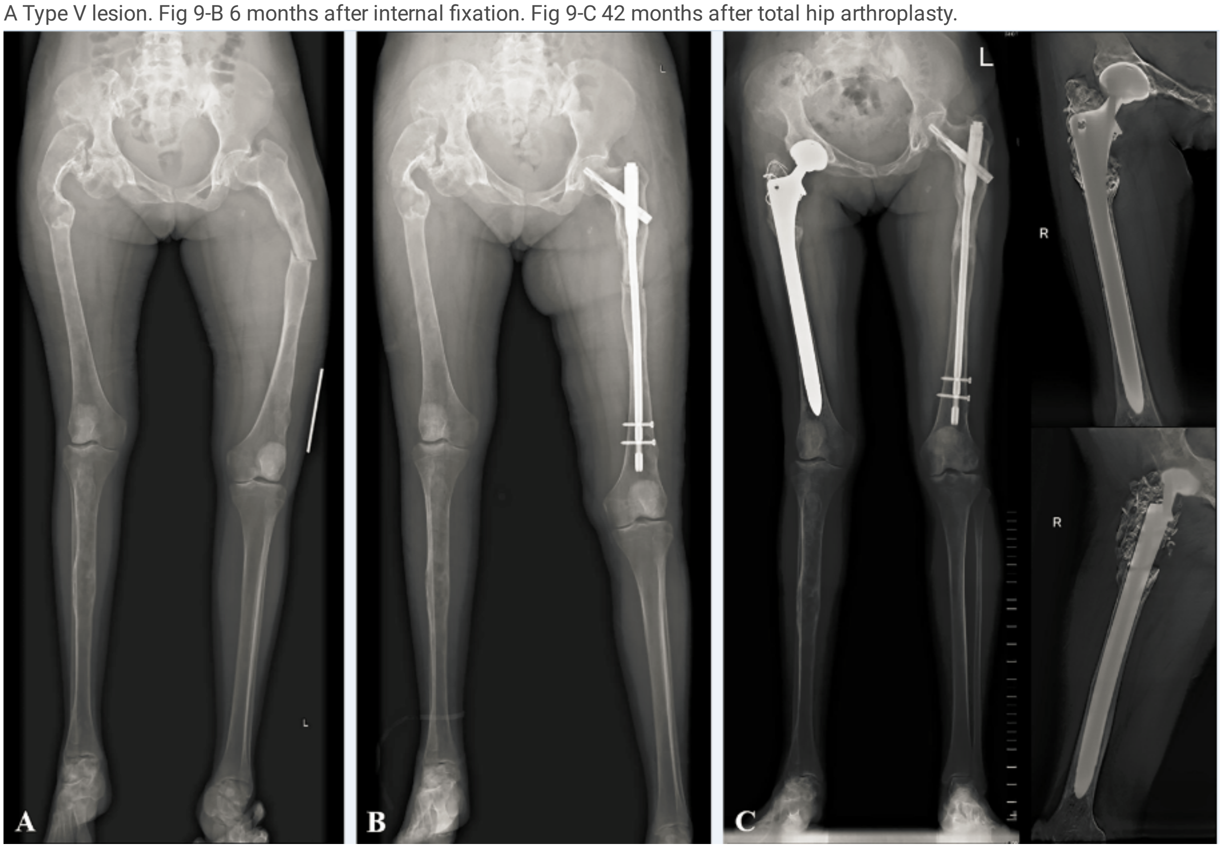


Figure 9

A Type V lesion. Fig 9-B 6 months after internal fixation. Fig 9-C 42 months after total hip arthroplasty.



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