Multiple osteotomies and intramedullary nailing with neck cross-pinning for shepherd’s crook deformity in polyostotic fibrous dysplasia

7 femurs with a minimum of 2 years follow-up

Sung-Taek Jung, Jae-Yoon Chung, Hyoung-Yeon Seo, Bong-Hyun Bae and Keun-Young Lim

Department of Orthopedic Surgery, Chonnam National University Hospital, Kwangju, Korea

Correspondence S-TJ: stjung@chonnam.ac.kr
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Background In polyostotic fibrous dysplasia, particularly in lesions of the proximal femur, pathological fracture and coxa vara deformity (including shepherd’s crook deformity) are likely to develop and progress.

Patients and methods In 7 femurs with shepherd’s crook deformity (5 patients), we performed intramedullary nailing by using multiple osteotomies and two screws crossing the femoral neck.

Results Restoration of the neck shaft angle of the femur was obtained from an average of 92º prior to surgery to 129º after surgery. During the minimum 2-year follow up, no loss of neck shaft angle of the femur and no refracture occurred. All patients were able to return to normal activities of daily living.

Interpretation In polyostotic fibrous dysplasia, multiple osteotomies and intramedullary nailing with neck cross-pinning can be used to correct developed or progressing shepherd’s crook deformity, and to prevent recurrence and refracture.

Patients and methods

From 2001 to 2003, 5 patients (7 femurs) with shepherd’s crook deformity received multiple osteotomies and M/DN (metaphyseal/diaphyseal nail) femoral interlocking nailing (Zimmer, Warsaw, IN). The average age was 24 (13–47) years, and there were 3 men (with 5 affected femurs) and 2 women (2 affected femurs). The average follow-up time was 30 (25–48) months. All patients experienced pain before surgery, and 4 patients had a his-
tory of previously healed fractures at the proximal femur. In 2 patients, to correct an accompanying deformity of the tibia, the tibia was osteotomized and nailed, and in 1 patient, to treat a limb-length discrepancy, shortening osteotomy and intramedullary nailing were performed on the contralateral femur (Figures 1 and 2).

The clinical outcome was assessed by a modification of the criteria described by Guille et al. (1998). Radiographically, union of the osteotomies, the neck shaft angle of the femur, limb length discrepancy, and mechanical axis deviation were compared before and after surgery, and cases with a difference in neck shaft angle of the femur of > 5° between immediately after surgery and at the final follow-up were considered to show loss of neck shaft angle.

The operation
The patient was placed in supine position under an imaging intensifier. Each preplanned osteotomy site was drawn on tracing paper, based on
Figure 2. Severe shepherd’s crook deformities of both proximal femurs in a polyostotic fibrous dysplasia patient (13-year-old boy). 3 years previously, he had undergone a simple nailing procedure at another hospital.
A, B and C. Preoperatively.
D, E and F. Immediate postoperatively.
G. At 2-year follow-up radiographs after corrective surgery with multiple osteotomies and intramedullary nailing with neck cross-pinning.
H and I. 1 year after correction of the femurs, both tibia were treated by intramedullary nailing.
preoperative radiographs. Intraoperatively, lateral closed-wedge osteotomy was performed where the cortical bone and guide pin were touching, with minimal exposure under an imaging intensifier. We confirmed passage of guide pin into the medullary canal through the open osteotomized site. During reaming, repeated removal of condensed fibrous tissues in the intramedullary cavity was required. After reaming, a proper nail was inserted. In all femurs, during surgery fibrous dysplasia was diagnosed by frozen biopsy, and the pathology was later confirmed by ordinary biopsy.

Postoperatively, a long leg splint was applied, and partial weight bearing was allowed approximately 2 weeks later. Walking time with full weight bearing was determined based on follow-up radiographical results.

Results

No infections, thromboembolism, or other notable complications occurred; however, in 1 femur, loosening of the distal locking screw of the intramedullary nailing was detected 4 months after surgery and the screw was removed. No additional loosening was detected at the final follow-up.

The clinical score according to the modified criteria of Guille et al. (1998) improved from an average of 3.6 (3–6) points prior to surgery to an average of 8.4 (6–10) points at the last follow-up. 5 patients achieved an excellent result, 1 was good, 1 fair, and no patient had a poor result (Table).

The neck shaft angle was corrected from average 92° (40–115) before surgery to average 129° (120–135) after surgery, and no change in the angle was detected at the final follow-up. Limb-length discrepancy was corrected from mean 27 (8–41) mm before surgery to mean 6 (2–11) mm after surgery. Mechanical axis deviation was corrected form mean 35 (5–45) mm medialization before surgery to mean 8 (5–15) mm medialization after surgery. In addition, all osteotomies healed at the final follow-up.

Discussion

In polystotic fibrous dysplasia patients with shepherd’s crook deformity, various surgical methods have been tried. Curettage of the lesion and bone grafting may be effective for mono-ostotic lesions (Freeman et al. 1987), but not for polyostotic fibrous dysplasia in the proximal femur (Funk and Wells 1973, Guille et al. 1998). In the shepherd’s crook deformity, Funk and Wells (1973) suggested the procedure of total excision of the diseased intertrochanteric area, and transplanted the insertions of the iliopsoas and the abductor muscles distal to the excised area to maintain function.

In cases of fracture or in those requiring correction for deformity, plate and screw fixation represents a relatively straightforward method. However, in the vicinity of weakened bones it is difficult to provide sufficient stability with screws, and fractures may easily occur due to the stress shielding effect of the distal part of the plate. Moreover, in the cases with a lesion over a wide area, due to a residual lesion, refractures may occur readily after metal plate removal (Guille et al. 1998). Also, in
cases with an advanced deformity, bowing deformity may also occur in the proximal and diaphyseal area; thus, in such cases requiring multiple osteotomies, it is inappropriate to use a plate and screw. In our study, 3 femurs had undergone deformity correction using a metal plate and screws at other hospitals, and lesion progression or refractures had occurred.

Connolly (1977) and Freeman et al. (1987) reported good results with multiple osteotomies and Zickel nail fixation for shepherd’s crook deformity. The advantage of this method is that it stabilizes the area of the lesion along the entire length of the femur, and by fixating the femoral head, the femoral neck is stabilized.

O’Sullivan and Zacharin (2002) reported that in 10 femurs (5 patients) with polyostotic fibrous dysplasia associated with McCune-Albright syndrome, bisphosphonate treatment and intramedullary nail fixation of the proximal femoral lesion prevented fractures and resulted in improved walking. However, the femoral neck was not stabilized, and loss of neck shaft angle occurred in 5 cases at follow-up. We experienced the same condition in 2 femurs before our operative procedure. Thus, we believe that stabilization of the femoral neck is important.

We did not use bisphosphonate therapy, but there have been some recent reports that bisphosphonate treatment of patients with polyostotic fibrous dysplasia reduces bone pain and risk of fracture, and improves walking ability (O’Sullivan and Zacharin 2002, Plotkin et al. 2003, DiCaprio and Enneking 2005).

We experienced some technical difficulties with our method. Firstly, due to the proximal varus deformity in the femur and muscle contracture between the greater trochanteric area and the ilium, the approach to the piriformis fossa for nail insertion was not easy. Secondly, as the first osteotomy area is the intertrochanteric or subtrochanteric area in most cases, the proximal part of the femur is short, and thus it is difficult to maintain the valgus position. However, this can be overcome by using the lateral decubitus position where the abductor muscles are extended by gravity. Thirdly, intramedullary reaming is made difficult by dense fibrous tissue.

Contributions of authors
STJ study conception and design, surgery, data analysis and writing the manuscript. JYC study conception and design, data analysis. HYS, BHB, KYL data analysis, interpretation and critical review of the manuscript.

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