
Case 92: Refracture, Soft Tissue Contracture, and Angular Deformity After Femoral Lengthening in Achondroplasia

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Contents

1	Brief Clinical History	653
2	Preoperative Clinical Photos and Radiographs	653
3	Preoperative Problem List	654
4	Treatment Strategy	654
5	Basic Principles	655
6	Images During Treatment	655
7	Technical Pearls	658
8	Outcome Clinical Photos and Radiographs	658
9	Avoiding and Managing Problems	658
10	See Also in Vol. 2	659
	References and Suggested Reading	659

Abstract

Extensive limb lengthening may be indicated in patients with achondroplasia in order to improve their quality of life. However, excessive lengthening is associated with decreased ROM of the adjacent joints, decreased new bone formation, delayed weight bearing, and fractures of the regenerate bones. Many surgeons have recommended that the goal of lengthening in a bone segment should be limited to 20 ~ 30 % of its initial length. And fractures of the regenerate bones may be avoided by careful analysis of the regenerate bone in the distraction gap before removal of the apparatus.

1 Brief Clinical History

Eight-year-old boy patient, who diagnosed with achondroplasia, was admitted for limb lengthening surgery. He underwent bilateral 8-cm lengthening of tibia 18 months ago. At the time of visit, he was complaining about gait abnormality caused by disproportionate limb length.

2 Preoperative Clinical Photos and Radiographs

See Figs. 1, 2, and 3.

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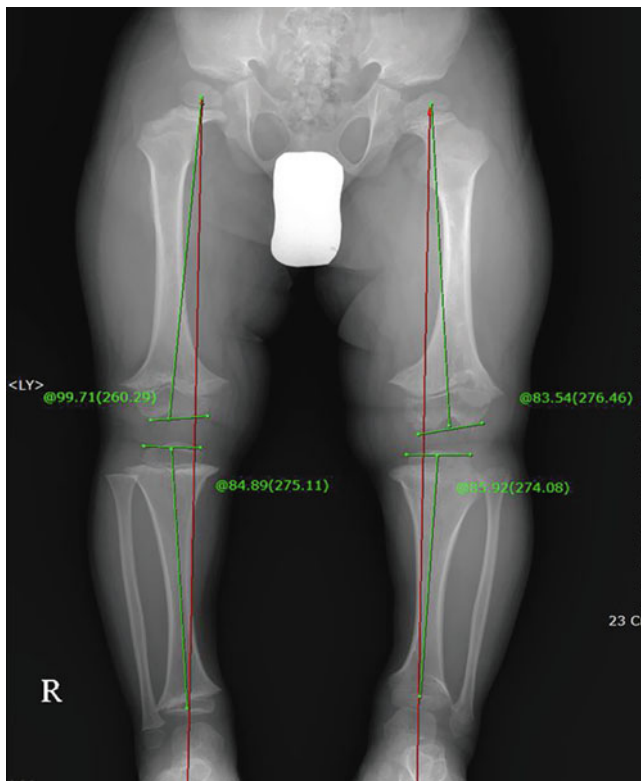


Fig. 1 Lower extremity AP at the time of initial visit (5 years 10 months). Rt.limb:mLDFA 99°, mMPTA 85°, MAD 13 mm. Lt.limb:mLDFA 84°, mMPTA 86°, MAD 14 mm



Fig. 2 Clinical photo of the patient

3 Preoperative Problem List

- I. Angular deformities
(Varus tendency at the lengthening site and valgus tendency of mechanical axis)
- II. Soft tissue contractures of the adjacent joints
(Flexion contracture of the hip, extension contracture of the knee, tightness of the iliotibial band)
- III. Poor callus formation and refracture of the regenerate bones
- IV. Difficulty in determining removal timing of the external fixators
(*Specific problems related with patients with achondroplasia*)
 - (i) Relatively short bone segment – The level of osteotomy should be located in mid-diaphyseal area.
 - (ii) Malalignment (coxa vara) of the proximal femur – Prone to fractures of the regenerate bone.
 - (iii) Previous tibial lengthening – Decreased potency of new bone formation in distraction osteogenesis and subsequent lateral instability of the knee joint.

4 Treatment Strategy

- I. Align the axis of external fixator according to the mechanical of axis of the lower extremity.
- II. Consider release of hip flexor muscles (sartorius, fascia lata, rectus femoris), quadricepsplasty for knee flexion, and Z-plasty of the iliotibial band for treatment of valgus knee.
- III. Avoid using wires for soft tissue impingement near the joint.
Consider half-pin insertion using the intermuscular planes.
- IV. Evaluate properly of the quality of regenerate bones.
Use the Pixel Value Ratio (PVR) method.
- V. Use PVR to determine proper timing of removal of the external fixators.
(*Specific treatment strategy related with patients with achondroplasia*)
 - (i) Timing of surgery – Consider significant disturbance of growth after extensive lengthening in skeletally immature patients with achondroplasia.

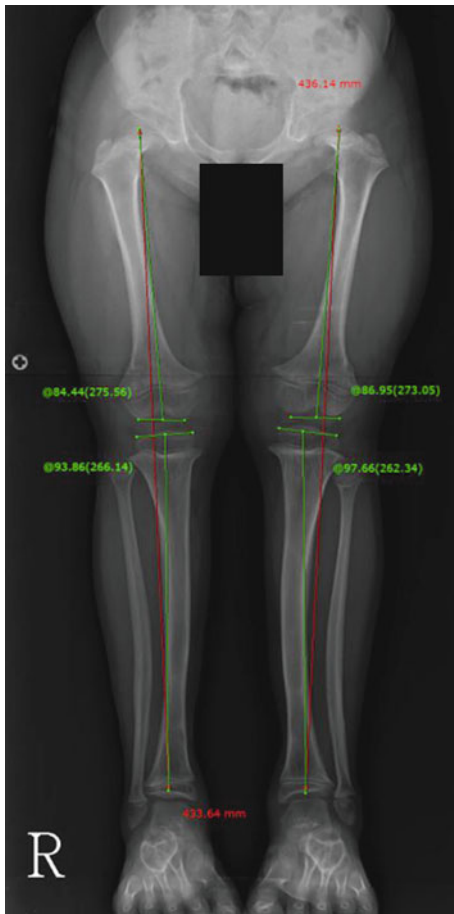


Fig. 3 POD 1 year 6 months after 8 cm of lengthening, correctional osteotomy, and external fixator application with Ilizarov apparatus (7 years 4 months). Rt.limb:mLDFA84°, mMPTA93°, MAD 10 mm. Lt.limb:mLDFA 87°, mMPTA 97°, MAD 14 mm

- (ii) Malalignment of the lower limb – Thorough preoperative evaluation of the patient's rotational profile and joint orientations.

5 Basic Principles

I. Determining the amount of lengthening

Excessive lengthening was associated with decreased new bone formation and delayed weight bearing. Large amount of lengthening more than 50 % of their initial femoral length was associated with an increased incidence of concave, lateral, and central callus shapes, adjacent joint stiffness, and fracture (Venkatesh et al. 2009).

II. Avoiding angular deformity

When an external fixator is used to lengthen the femur, the device can be aligned with the mechanical or the

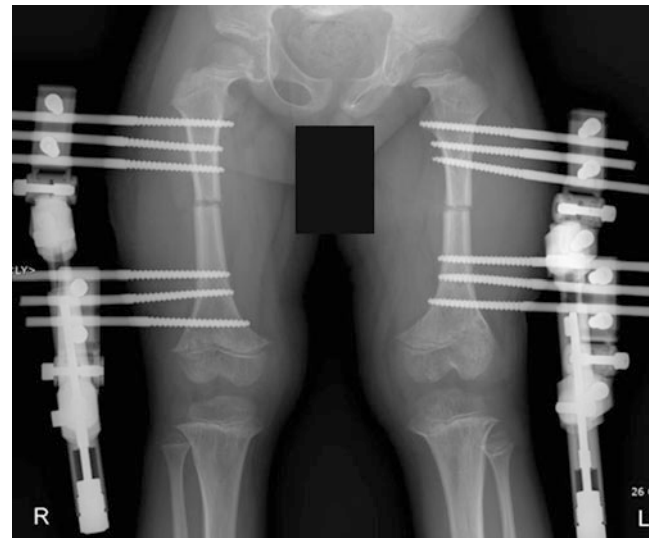


Fig. 4 After femoral lengthening osteotomy and external fixation with monolateral fixator (8 years 2 months)

anatomical axis. Aligning it with the anatomical axis theoretically leads to medialization of the knee joint (lateralization of the mechanical axis line). Therefore, most femoral external fixators should be aligned with the mechanical axis.

III. Management of soft tissue

Excessive lengthening is associated with a decreased ROM of the adjacent joints. Knee movement is known to help to prevent muscle atrophy and adhesion of the muscles to the underlying bone regenerate and to increase the blood supply to bone and bone strength (Herzenberg et al. 1994). Excessive lengthening affects the thigh muscles by unable to keep pace with the lengthening, resulting in a decreased ROM at the knee and fixed flexion at the hip.

IV. Evaluation of the regenerate bone (Pixel Value Ratio, PVR)

The pixel values of the proximal, distal, and regeneration areas could be calculated from the mean value of each area using the free line range of interest (ROI) method.

V. Timing of the external fixator removal (PVR)

A pixel value ratio of 1 indicated corticalization of the regenerate in the lengthening area. The external fixators should be removed based on a PVR of at least 1.0.

6 Images During Treatment

Figs. 4, 5, 6, 7, 8, 9, and 10.

Fig. 5 POD 3 months after 9 cm of lengthening (8 years 5 months) and Assessment of Pixel value based on a radiograph in PACS system. Note the red circle showing the pixel value of range of interest (*ROI*) drawn at each segment. The value *M* (encircled in red) indicates the mean pixel value for that segment and can be used in the computation for the PVR pixel value ratio = [(pixel value of the proximal segment + pixel value of the distal segment) / 2] / pixel value of the regenerate (Song et al. 2011)

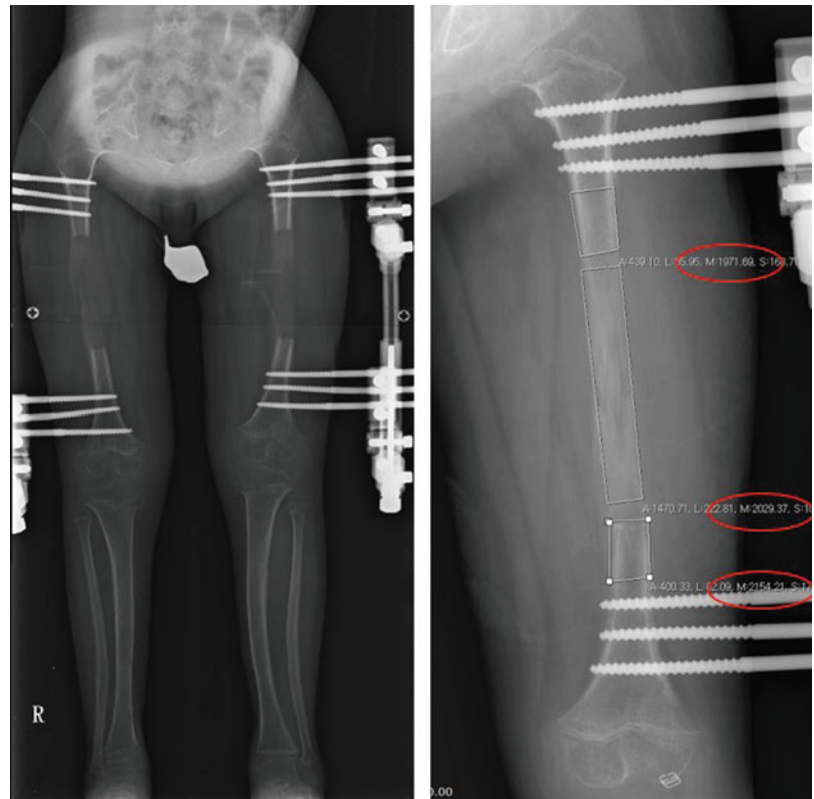


Fig. 6 Clinical photos showing flexion contracture of the hip joint caused by excessive lengthening. He was treated with releasing of sartorius, fascia lata, straight head of rectus femoris, and psoas muscle

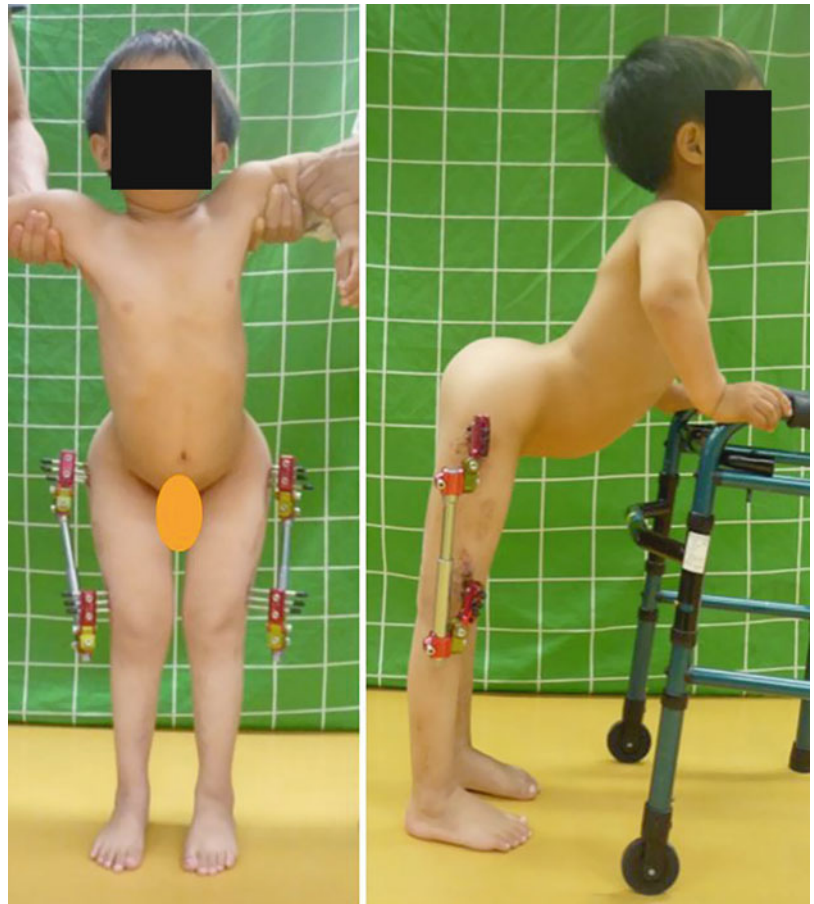




Fig. 7 POD 1 year 3 months after 9 cm of lengthening (9 years 5 months) showing concave and lateral type of poor callus formation

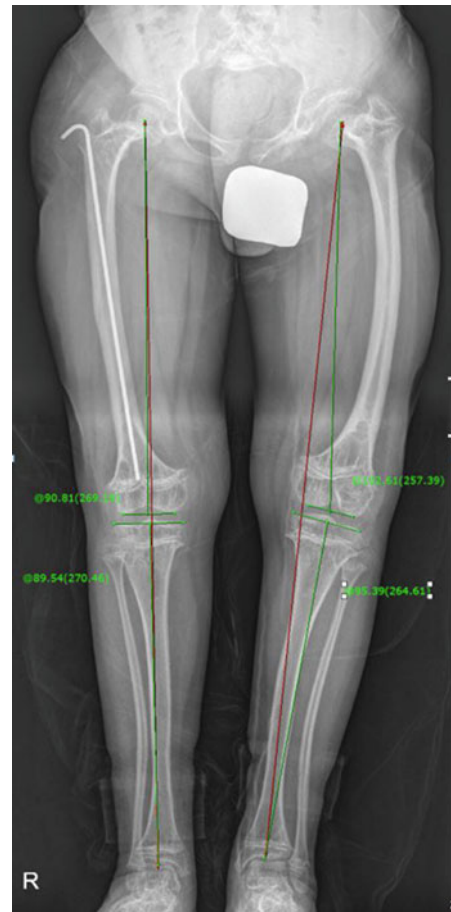


Fig. 9 POD 1 year (11 years 5 months) after removal of implant on left side. Rt.limb:mL DFA91°, mMPTA89°, MAD 2 mm. Lt.limb: mL DFA102°, mMPTA95°, MAD 19 mm

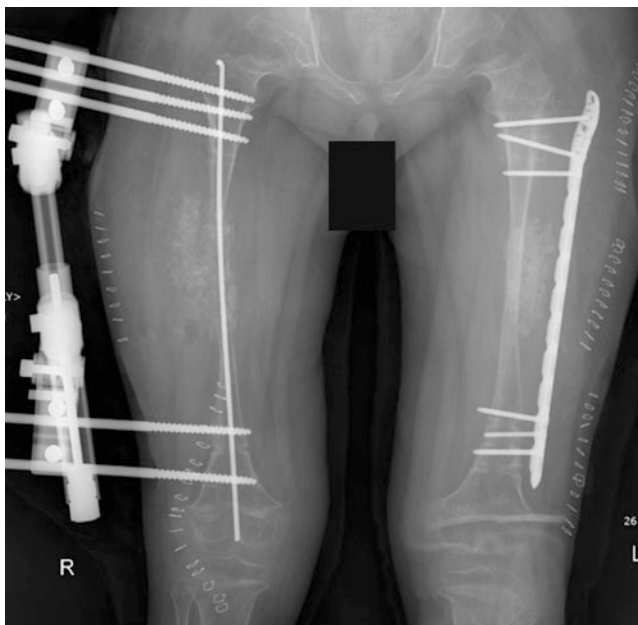


Fig. 8 Third operation for regenerate fracture with additional bone graft (DBM and PRP) and internal fixation (9 years 5 months)

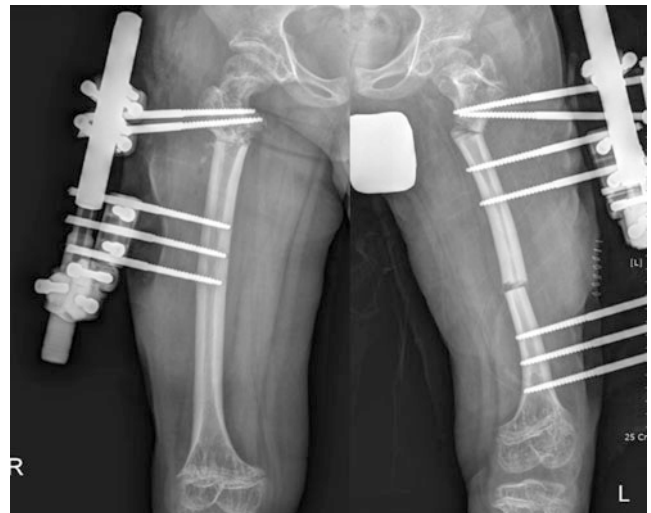


Fig. 10 After bifocal femoral osteotomy and monolateral external fixator application on left limb (11 years 5 months) and monofocal femoral osteotomy and monolateral fixator application on right limb (12 years 4 months)

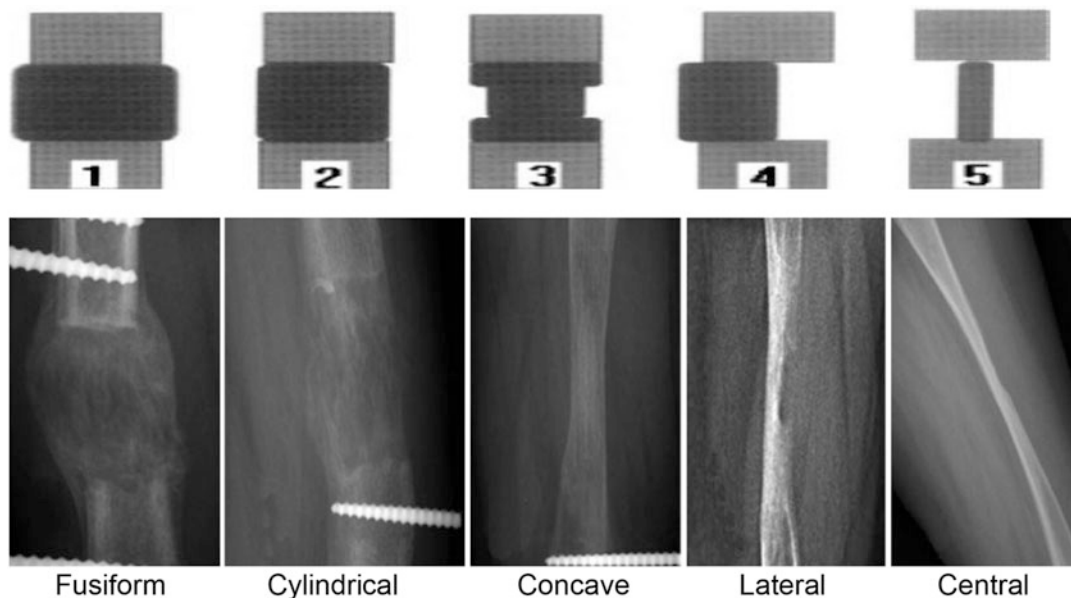


Fig. 11

7 Technical Pearls

- I. Early physal closure related to lengthening loss ranging from 1.5 to 4 cm after extensive tibial and femoral lengthening could occur after extensive limb lengthening in skeletally immature patients with achondroplasia (Song et al. 2012) and start lengthening after 10 years old.
- II. Aligning the external fixators with the mechanical axis did not guarantee anatomical alignment after lengthening. There is tendency of varus angulation due to absence of medial distraction when using monofixator and tendency of valgus knee caused by iliotibial band.
- III. Regular radiological examination, physiotherapy, careful adjustment of the distraction rate, and soft tissue releases should be considered during large amount of femoral lengthening. And regular stretching and the maintenance of a good ROM can help to decrease the pain and muscle spasm during lengthening.
- IV. The radiological features of the callus in distraction osteogenesis could be classified with regard to shape and type on the basis of the Li classification (Li et al. 2006). The shape was based on the width of the callus compared to the original osteotomy site. The type was based on the callus shapes (cylindrical, fusiform, and concave), three patterns of osteogenesis (homogeneous, heterogeneous, and lucent), and three densities (low, intermediate, and normal). When there is concave or lateral type of poor

callus formation, compression is necessary until fusiform or cylinder type of callus is visible (Fig. 11).

Description	Name
Regeneration wider than the original bone	Fusiform
Regeneration same width as the original bone	Cylindrical
Regeneration narrower than the original bone	Concave
Regeneration mainly on one side of the distraction gap	Lateral
Regeneration a thin pillar	Central

- V. We can safely rely on PVR values to define the timing of fixator removal, as it exhibits a good correlation in patients with a cylindrical callus pattern regardless of whether the pathway is homogeneous or heterogeneous (Song et al. 2011).

8 Outcome Clinical Photos and Radiographs

See Figs. 12 and 13.

9 Avoiding and Managing Problems

- I. Treating physicians should include the possibility of early physal closure in preoperative counseling of the patients and their parents and also should be cautious in determining the amount of lengthening.

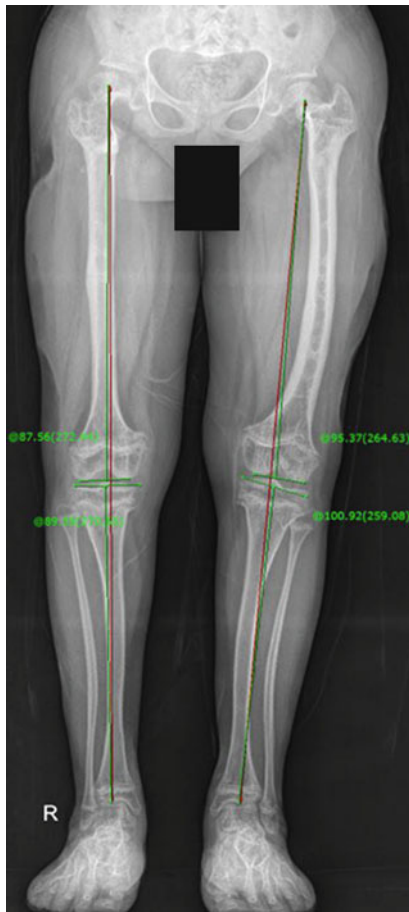


Fig. 12 POD 6 months after femoral correction (13 years). Rt.limb: mLDFA87°, mMPTA89°, MAD 2 mm. Lt.limb: mLDFA95°, mMPTA100°, MAD 2 mm



Fig. 13 Clinical photo at the time of last visit (13 years)

- II. Treating physicians should consider difference of axis between mechanical axis and anatomical axis in coronal plane and axial deviation during femoral lengthening and also their surrounding soft tissue conditions.
- III. Various soft tissue procedures such as release of the fascia of vastus lateralis, tensor fascia lata, psoas tendon, hamstring tendon, and iliotibial band could be used to correct joint stiffness.

10 See Also in Vol. 2

Case 8: Femoral Bone Defect

References and Suggested Reading

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