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journal or publication title	Journal of Orthopaedic Science
volume	11
number	5
page range	459-466
year	2006-09-01
URL	http://hdl.handle.net/2297/2956

Correction and lengthening for deformities of the forearm in multiple cartilaginous exostoses

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Running title: Correction of forearm exostoses

Abstract

Background. Multiple cartilaginous exostoses cause various deformities of the epiphysis. In exostoses of the ulna, it is shortened and the radius acquires varus deformity. Sometimes this leads to dislocation of the radial head. In this study, we present the results of exostoses resection, and correction and lengthening with external fixators for functional and cosmetic improvement, and prevention of radial head dislocation.

Methods. We retrospectively reviewed seven forearms of seven patients who had deformities of the forearm associated with multiple cartilaginous exostoses. One patient had dislocation of the radial head. Operative technique was excision of osteochondromas from the distal ulna, correction of the radius, and ulnar lengthening with external fixation up to 5 mm plus variance. We evaluated radiographs and the range of pronation and supination. Furthermore we conducted a follow-up of the ulnar length after the operation.

Results. Dislocation of the radial head of one patient was naturally reduced without any operative intervention. At the most recent follow-up all patients except one showed full improvement in pronation-supination. Ulnar shortening recurred with skeletal growth of four skeletally immature patients; however, it did not recur in one skeletally mature patient. Overlength of 5 mm was negated by the recurrence of ulnar shortening about one-and-a-half years after the operation.

Conclusions. We treated seven forearms of seven patients by excision of osteochondromas, correction of radii, and gradual lengthening of ulnas with external fixators. The results of the procedure were satisfactory, especially for the function of the elbow and wrist. However, we must consider the possible recurrence of ulnar shortening within about one-and-a-half years during skeletal growth periods in immature patients.

「多発性外骨腫症による前腕変形短縮の創外固定器による治療経験」

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【目的】多発性軟骨性外骨腫症による尺骨の変形・短縮は、橈骨の彎曲変形やそれに伴う橈骨頭脱臼などを合併し、整容的のみならず機能的にも障害をきたす。我々は整容的な改善と橈骨頭脱臼の予防のために外骨腫切除とともに創外固定器を用いて矯正延長を行ったので報告する。

【方法】多発性軟骨性外骨腫症による前腕変形7例7肢(男3女4例)に対して手術を施行した。手術時の平均年齢は10.8歳(5~13)、平均経過観察期間は7.1年(2.7~14.1年)であった。全例、尺骨遠位端に外骨腫を形成し、短縮を伴っていた。尺骨の外骨腫を切除後に、橈骨は一次的に内反変形を矯正し内固定を行なうか、創外固定器を設置して緩徐に矯正を行った。尺骨は創外固定器を用いて仮骨延長を行なった。尺骨の延長は、相対的短縮の再発を考慮し、尺骨・橈骨の等長よりも延長終了時で、5mm前後の過延長を目標とした。

【結果】術前、尺骨は橈骨に対して平均12.3mm(10~15mm)短縮しており、橈骨は健側と比し平均17度内反変形していた。尺骨の延長は平均27mm施行した。成長に伴う変形の再発のため1例が初回手術から2年3ヵ月後に再手術を要した。尺骨の相対的延長量は約1.5年で相殺された。橈骨頭脱臼を認めた1例は、尺骨の延長に伴い自然と整復された。前腕の回内外は術前には2例で制限を認めており、矯正終了時で5例に回内外制限を認めた。最終評価時には1例で回内制限を認めたが日常生活上は問題なかった。

【考察】尺骨の相対的短縮の再発を考慮すると、尺骨の過延長を行なうべきである。一次的延長では20mmが限界といわれており、症例によっては尺骨の過延長を行えなくなる。しかし創外固定器を用い緩徐に延長することで、神経血行障害が回避しやすく、また短縮した橈骨の延長も可能である。また、橈骨頭が脱臼している症例であっても創外固定器で延長を行いながら、橈骨頭の脱臼を整復させることが可能である。

【結論】創外固定を用いた多発性軟骨性外骨腫症による前腕変形短縮に対する治療は、治療期間は長くなるが、有用な方法となりうる。しかしながら、約1.5年で尺骨の相対的延長量は相殺されることを念頭において手術を考慮すべきである。

Introduction

Multiple cartilaginous exostoses are a hereditary condition, in which exostoses are formed around the joints and cause various deformities of the epiphysis. Exostoses of the forearm are mostly formed in the distal ulna, which lead to growth disturbance. While growing, the ulna is shortened and the radius acquires varus deformity. Sometimes this leads to dislocation of the radial head. Some cases undergo malignant transformation and become secondary chondrosarcomas.¹

Although various treatment alternatives have been proposed to deal with such deformities and shortening of the forearm, lengthening of the ulna with correction only or with lengthening of the radius is considered to be the most appropriate treatment. However, most reports show only short term results and little is known about the postoperative course. In this study, we present the results of resection of exostoses, and correction and lengthening with external fixators for the purpose of functional and cosmetic improvement, and prevention of radial head dislocation.

Patients

We retrospectively reviewed seven forearms of seven patients who had deformities of the forearm associated with multiple cartilaginous exostoses (Table 1). The patients were treated in our hospital between 1991 and 2002. There were 3 boys and 4 girls, ranging in age from 6.8 to 14 years (average 10.8 years). The follow-up period ranged from 2.7 years to 14.1 years (average 7.1 years). There was deformation in 3 right and 4 left forearms, dislocation of radial head was found in one patient, and there were two patients who had a restriction of pronation before surgery. An Ilizarov fixator was used alone in one patient, monotube fixation in three patients, both in one patient, and Orthofix (M-100) in two patients. A unilateral fixator was selected when angulation of the radius could be safely corrected in an acute manner, followed by gradual lengthening of the ulna. Monotube or Orthofix (M-100) fixators were selected for their fit of the size of the ulna. The Ilizarov external fixator was used in cases that required gradual multifocal or oblique plane corrections or lengthening of the radius.

Surgical indications included radial head subluxation-dislocation, disturbance of activities of daily living due to limited or painful forearm rotation, progressive deformity, and undesirable cosmetic appearance.

Operative technique

The operative technique was as follows (Fig. 1).

(1) Excision of osteochondromas of the distal ulna, (2) corrective osteotomy and internal fixation of the radius (5 patients) or osteotomy and gradual correction of the radius with an

external fixator (2 patients), (3) gradual lengthening of the ulna using an external fixator up to 5 mm plus variance, with the expectation of subsequent recurrence of ulnar shortening.

Evaluation

Radiographic review compared preoperative and most recent follow-up radiographs. We measured the radial articular angle (RAA; Fig. 2A), bowing of the radius, the carpal slip (CS; Fig. 2B), and the relative ulnar shortening (Fig. 2C).² We also evaluated the amount of ulnar lengthening, correction angle of the radius, external fixation time (EFT), external fixation index (EFI), and the distraction index (DI). EFI was obtained by dividing the total duration of external fixation by the length gained; DI was obtained by dividing the total duration of distraction by the length gained. We evaluated the range of pronation and supination clinically. Furthermore we conducted a follow-up of the ulnar length after the operation.

Results

Before the operation, the ulna was shortened by an average of 12.3 mm (range 10 to 15 mm), resulting in a bowing of the radius by an average of 21.9 degrees (range 18 to 27 degrees). RAA before surgery was an average of 38.6 degrees (range 30 to 49 degrees). CS before surgery was an average of 56.7% (range 29 to 100 %).

At the most recent follow-up, RAA was an average of 25.7 degrees (range 15 to 36 degrees), improved compared to the normal side. The ulna was gradually lengthened by an average of 27 mm (range 16 to 41 mm), resulting in 1.7 mm ulnar length minus variance at the most recent follow-up. However, in four patients shortening of the ulna recurred, and in one patient 13 mm re-lengthening of the ulna was performed. CS was improved by about 16%. Correction angle of the radius was an average of 16 degrees (range 0 to 25 degrees) (Table 1). EFT was an average of 126 days, EFI was an average of 52.9 days/cm, and DI was an average of 16.0 days/cm. Dislocation of the radial head of one patient was naturally reduced without any operative intervention. Before surgery two patients had a restriction of pronation; however, five patients had a restriction of pronation-supination just after completing the lengthening. That is because of the ulno-carpal impaction syndrome. At the most recent follow-up, six patients showed full improvement in pronation-supination. The remaining patient had a slight restriction of supination. All of the tumors resected were pathologically confirmed as exostoses and none were malignant. No tumor recurrence was found during the follow-up period. Superficial infection occurred in one patient and was treated with antibiotics, but there was no deep infection, or breakage of wires or pins.

Ulnar shortening recurred with growth of four immature patients; however, it did not recur in one skeletally mature patient. Overlength of 5 mm was negated by the recurrence of ulnar

shortening within about one-and-a-half years of the operation (Fig. 3).

Case presentations

Case 2 (Fig. 4)

A 14-year-old boy complained of deformity of the left forearm and limitation of pronation due to a giant exostosis. Range of pronation was 30 degrees. On X-ray RAA was 33 degrees, CS was 66%, and ulnar shortening was 12 mm (Fig. 4A, B). During the operation, the range of pronation improved to its full extent after resecting an exostosis of the distal ulna. The radius was corrected acutely and fixed with a plate, an Orthofix (M-100) external fixator applied to the ulna, and osteotomy performed (Fig.4C). The ulna was lengthened 22 mm after a waiting period of 10 days. External fixation time was 141 days. At the most recent follow-up, he had no pain, unlimited range of pronation, and no recurrence of ulna shortening (Fig. 4D, E).

Case 6 (Fig. 5)

An 11-year-old girl complained of deformity of the right forearm and limitation of pronation. Range of pronation was 30 degrees. X-ray radiography demonstrated that RAA was 33 degrees, CS was 66%, ulnar shortening was 12 mm, and lateral dislocation of the radial head (Fig. 5A–C). The radius was corrected acutely and fixed with a plate, a monotube external fixator applied to the ulna, and osteotomy performed. The ulna was lengthened 45 mm after a waiting period of 7 days. The radial head was reduced without any open reduction for dislocated radial head. External fixation time was 155 days (Fig. 5D). At 3 years 8 months after the operation, she had no pain and unlimited range of pronation. There was no hindrance to her activities of daily living. Furthermore, on X-ray radiographs, no recurrence of ulnar shortening and dislocation of radial head could be seen (Fig. 5E, F).

Discussion

Multiple cartilaginous exostoses are a benign autosomal dominant skeletal dysplasia affecting bones formed by endochondral ossification. About 40% of the patients are sporadic. Estimations of its prevalence have ranged from between 0.9 and 1.4 in 100,000,³ in two European populations. There has been a wide variation in the reported rates of malignant degeneration of a benign exostosis to chondrosarcoma or to some other sarcoma in patients who have hereditary multiple cartilaginous exostoses. Reports have documented the risk to be 2 to 5 percent.⁴ Multiple cartilaginous exostoses often cause an obvious deformity of the forearm. The incidence of forearm lesion in multiple exostosis patients was reported to be between 39 and 59%.^{3,5}

Prichett reported that the characteristic relative ulnar shortening is related to three factors.¹

1. The ulnar physis had a cross-sectional area that is only one-quarter of that of the distal radius.
2. The distal ulna is more commonly involved in the condition than the distal radius.
3. The distal ulnar physis contributes more to total ulnar length than the distal radial physis contributes to radial length.

Osteochondroma in the distal ulna can result in a relative shortening of the ulna called candy stick deformity, increased inclination of the distal radial articular surface with ulnar migration of the carpus, and sometimes proximal radial head subluxation or dislocation.⁶

For these deformities, several surgical interventions have been reported.⁷⁻¹¹ Fogel reported three different procedures,⁷ excision of osteochondromas only, ulnar lengthening with excision of osteochondroma, and ulnar lengthening with radial hemiepiphyseal stapling and excision of osteochondromas. Excision of distal osteochondromas did not improve the deformity, but it did reduce the preoperative rate of progression of ulnar shortening. Ulnar lengthening with excision of osteochondroma did not result in a significant improvement in forearm rotation, radial articular angle, or carpal slip. On the other hand, acute ulnar lengthening (0.3–1.6 cm) with radial hemiepiphyseal stapling and excision of osteochondromas led to good results. However, this procedure made the final length discrepancy unpredictable as the final length of the forearm could not be predicted.

Wood reported other procedures, closing wedge osteotomy of the radius and fixing with a compression plate and screws, excision of the osteochondroma of the distal ulna, and ulnar lengthening using a long stepcut osteotomy and securing the bone in its lengthened position. Although appearance was markedly improved using this method, functional improvement was only minimal.¹⁰

Prichett reported that lengthening of the ulna and correction of the radius, particularly with the use of an external fixator, has given predictable results and is a useful method of treating forearm deformity in patients with hereditary multiple exostoses.¹

It appears that we reached the consensus of a need for lengthening and normalization of the relationship of the radius and ulna, but controversy in the literature now exists regarding whether forearm lengthening is best achieved immediately or gradually. Some authors have reported that immediate lengthening of the ulna should be limited to 20 mm because greater lengthening can cause neurovascular problems.^{6,12} On the other hand, Waters reported that it was safe to lengthen acutely up to 25 mm or 20% of total length.¹³

To prevent possible recurrence, however, it would be better to overlengthen the shorter ulna. Most patients required lengthening from about 20 to 40 mm considering the recurrence.¹⁵ By lengthening gradually, patients can avoid problems of the neurovascular system and lengthen

the shorter radius as well. Stapling or hemi-epiphysiodesis of the distal radial physis, or shortening of the radius to prevent recurrence were reported by Siffert.¹⁵ This is not acceptable because the length of affected limbs will be shorter. Furthermore, a dislocation of the radial head can be reduced using gradual lengthening with an external fixator.¹ Therefore we believe that our method is a good way to treat forearm deformity due to osteochondroma. The timing of the surgery is extremely important, and there are many reports regarding this issue. Some recommend early intervention, because earlier intervention has more potential for remodeling and leads to better surgical results.^{1,8,9,16,17} Others recommend intervention at a later age, because a recurrent operation can be avoided by postponing the procedure and good function can be acquired despite significant deformity after skeletal maturity.^{14,18} We perform the operation if the patients have the indication without considering their age. This is because some patients lose function which can not be regained by postponing the operation, especially with a dislocation of the radial head.

There is some literature about follow-up of lower leg lengthening,^{19,20,21} and there is a smaller body of literature regarding the middle- to long-term follow-up of ulnar lengthening. In this study, our results showed that an overlength of 5 mm was negated by the recurrence of ulnar shortening about one-and-a-half years after the operation. Monte reported that the tendency for recurrence of ulnar shortening was inversely proportional to the age at operation. It varied from 5.3 mm per year in patients aged five years, to 2.7 mm per year in patients aged eleven years.⁶ This finding is similar to ours, and provides surgeons with useful information to explain the operation to the patients and their parents. However, our results demonstrated no relationship between the amount of recurrent ulnar shortening and patient age at operation. We hypothesize that the amount of recurrent ulnar shortening depends on the extent of damage of the distal ulnar growth plate due to the osteochondroma.

Conclusion

We treated seven forearms of seven patients by excision of osteochondromas, correction of the radius, and gradual lengthening of the ulna with an external fixator. The results of the procedure were satisfactory, especially for the function of the elbow and wrist. However, we must consider the possible recurrence of ulnar shortening within about one-and-a-half years during skeletal growth periods in immature patients.

The patients and their families were informed that data from the case would be submitted for publication, and gave their consent.

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Figure legends

Fig. 1. Operative technique

- A. Excision of osteochondromas of distal ulna
- B. Osteotomy
- C. Correction of the radius, gradual lengthening of the ulna

Fig. 2. Evaluation on radiographs

- A. Radial Articular Angle (RAA) is the angle between two constructed lines : one along the articular surface of the radius and the other perpendicular to a line that bisects the head of the radius and passes through the radial edge of the distal radial epiphysis.
- B. Carpal Slip (CS) is measured as the percentage of contact of the lunate with the radius, determined by an axial line drawn from the center of the olecranon through the ulnar edge of the radius.
- C. Ulnar shortening (a) is measured with a perpendicular drawn from the distal end of the ulna to the linear axis of the forearm.

Fig. 3. Sequence of ulnar shortening

Fig. 4. Case 2 (14-year-old boy)

- A. Restriction of pronation before the operation
- B. Radiographs before the operation (front and lateral view)
- C. Radiographs after the operation (front and lateral view)
- D. Radiographs at 1.8 years after the operation (front and lateral view)
- E. Full range of pronation and supination at 1.8 years after the operation

Fig. 5. Case 6 (11 year-old girl)

- A. Restriction of pronation before the operation
- B. Deformity and shortening of her right forearm
- C. Radiographs before the operation (front and lateral view), arrow: dislocation of radial head
- D. Radiographs after the removal of the external fixator (front and lateral view): Radial head was reduced naturally (arrow)
- E. Radiographs at 3.7 years after the operation (front and lateral view)
- F. Full range of pronation and supination at 3.7 years after the operation

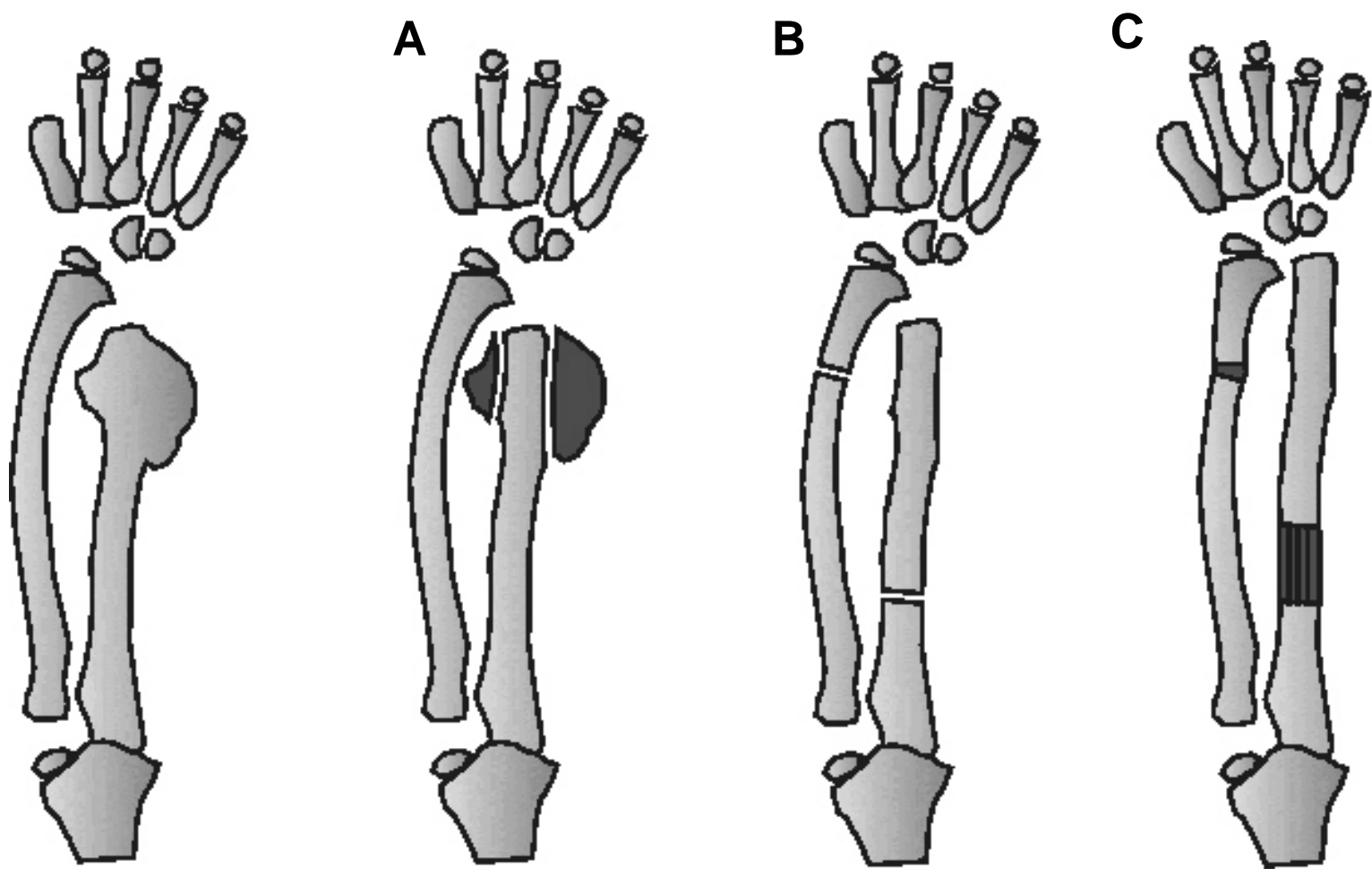


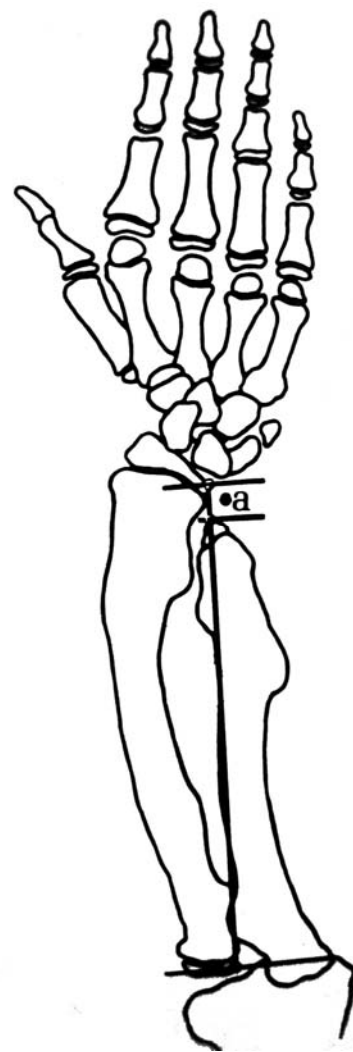
Fig.1



A



B



C

Fig.2

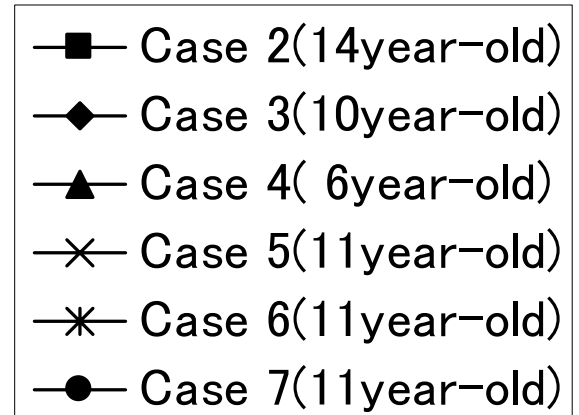
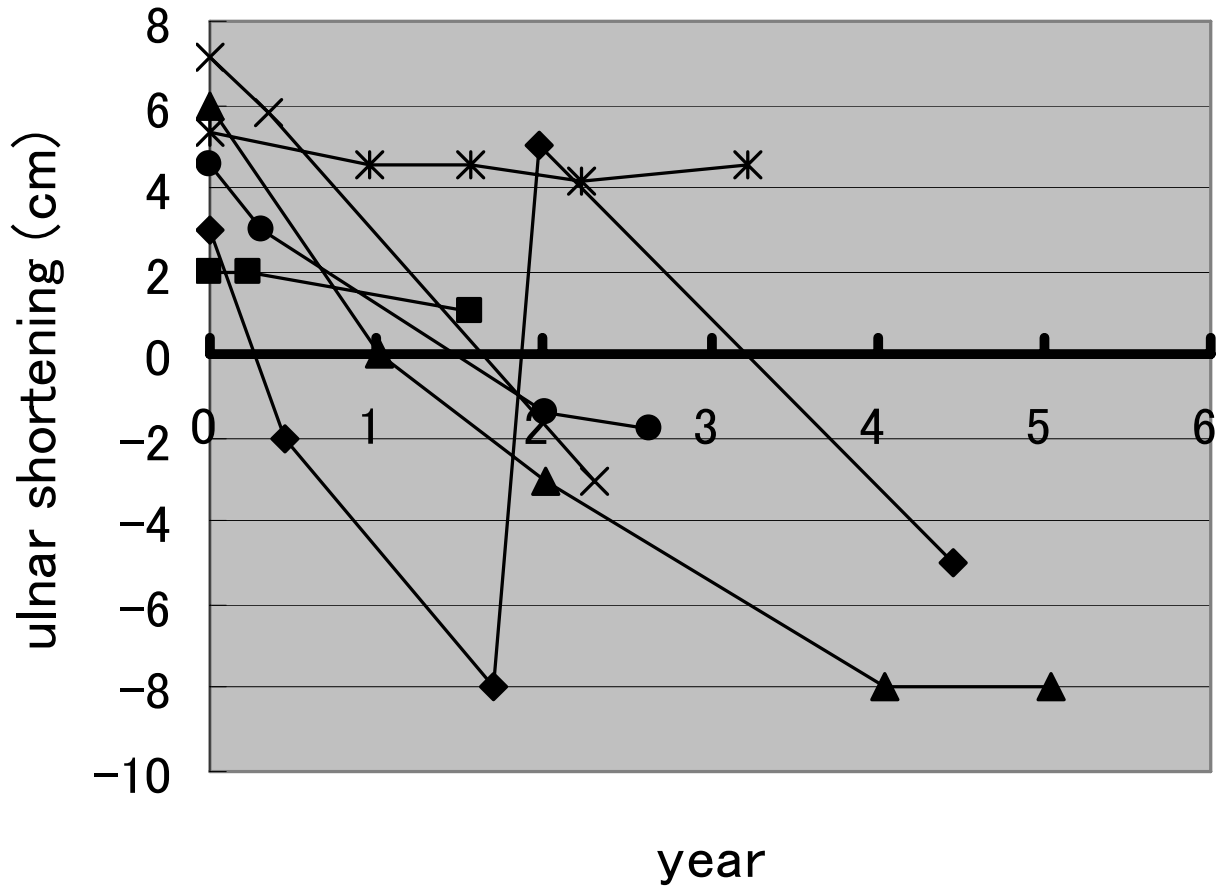


Fig.3

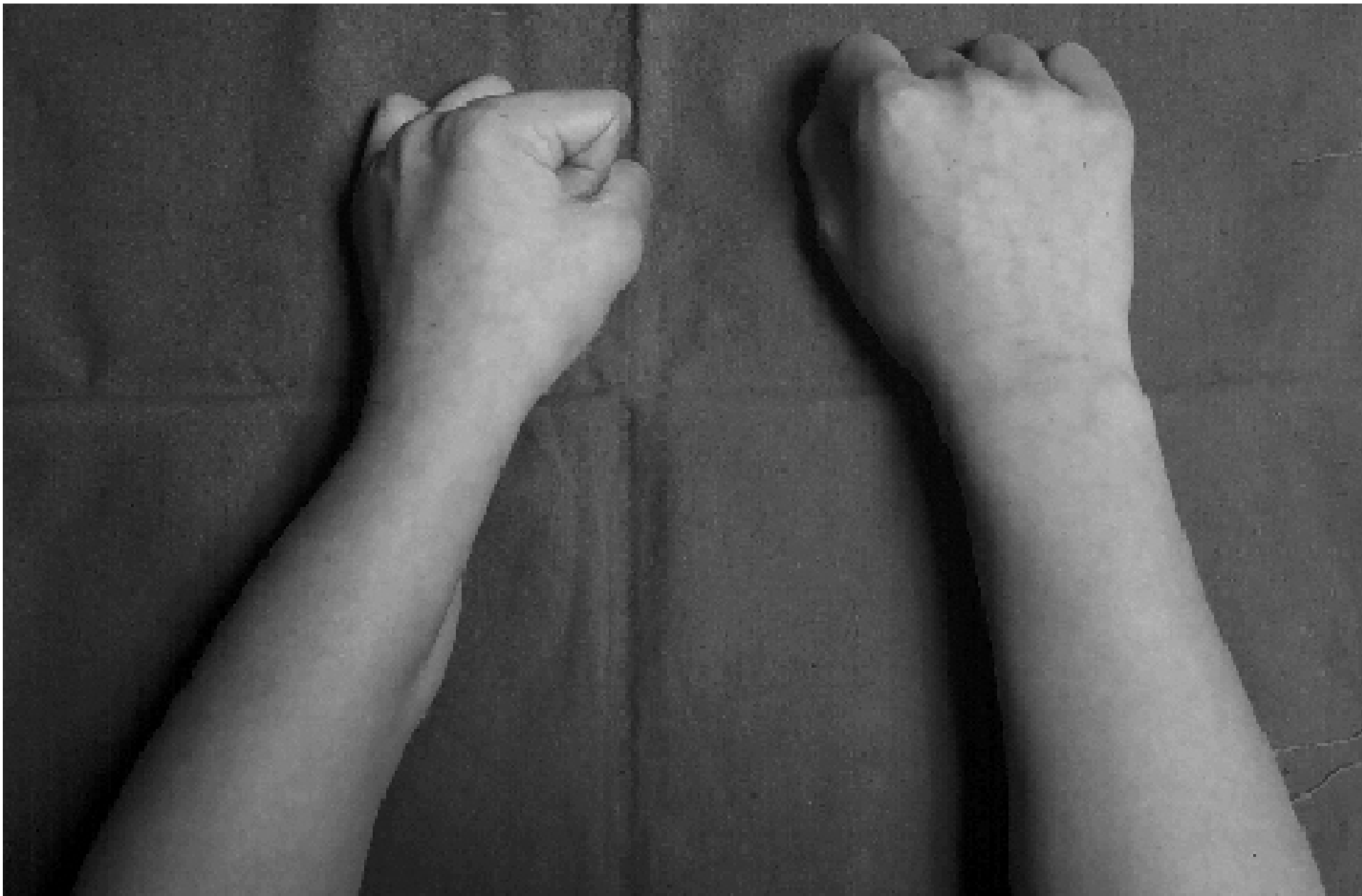


Fig.4A



Fig.4B

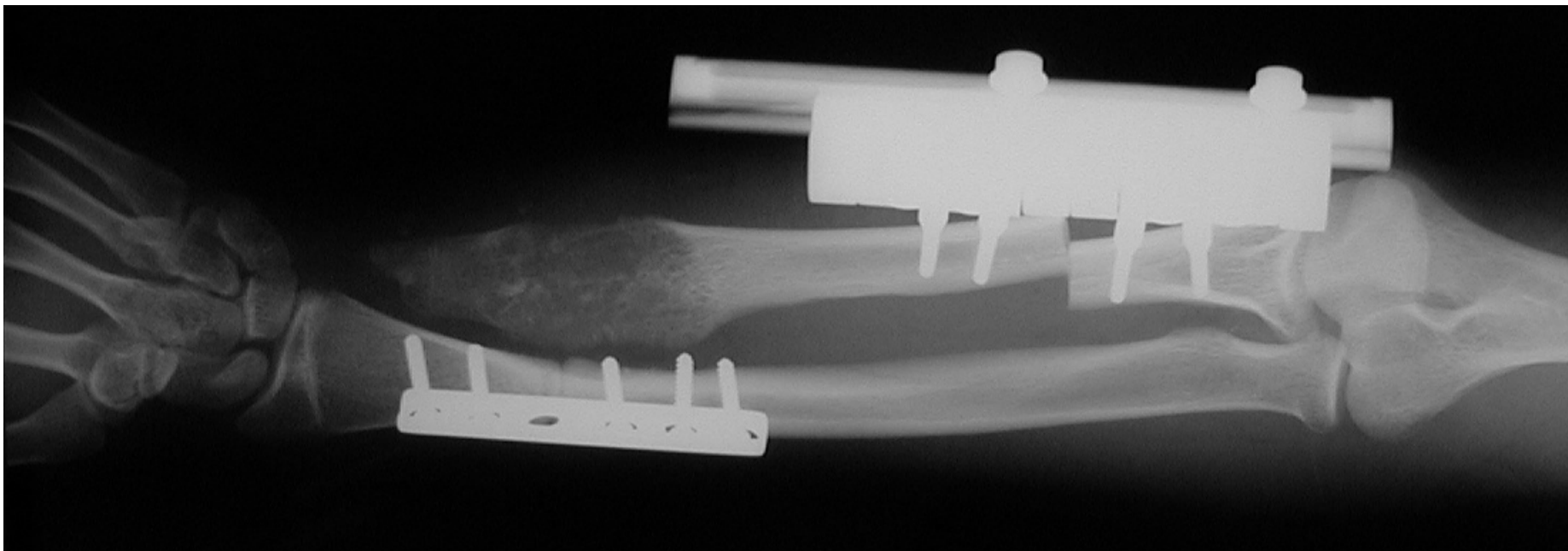
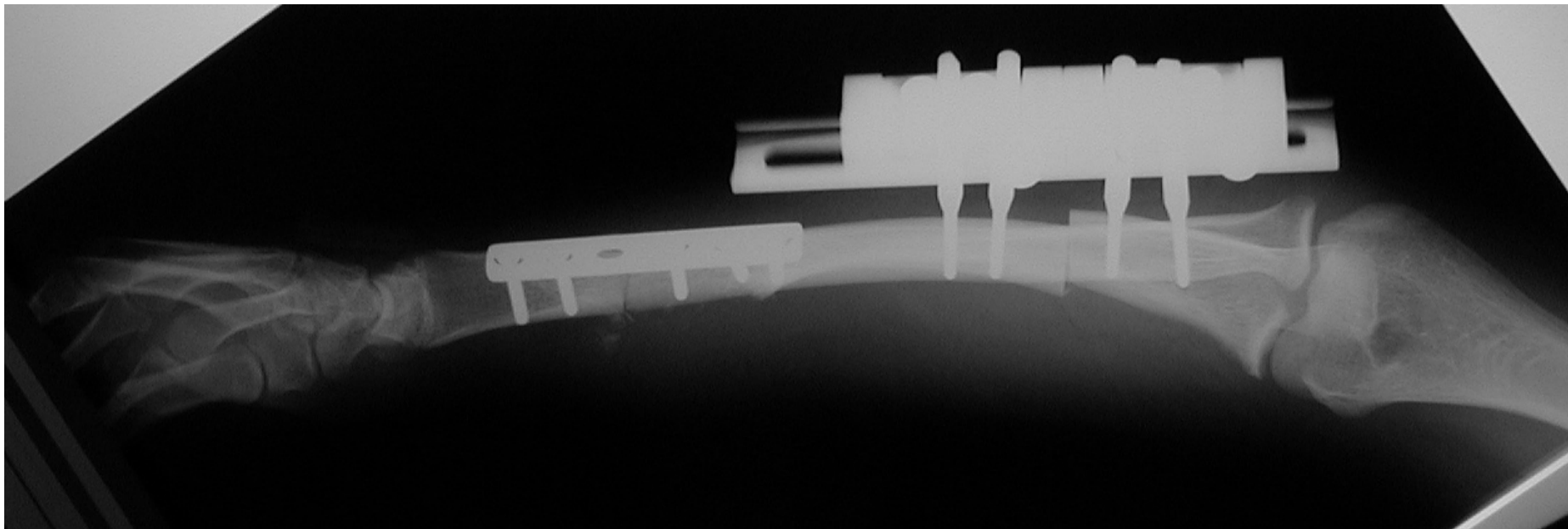


Fig.4C

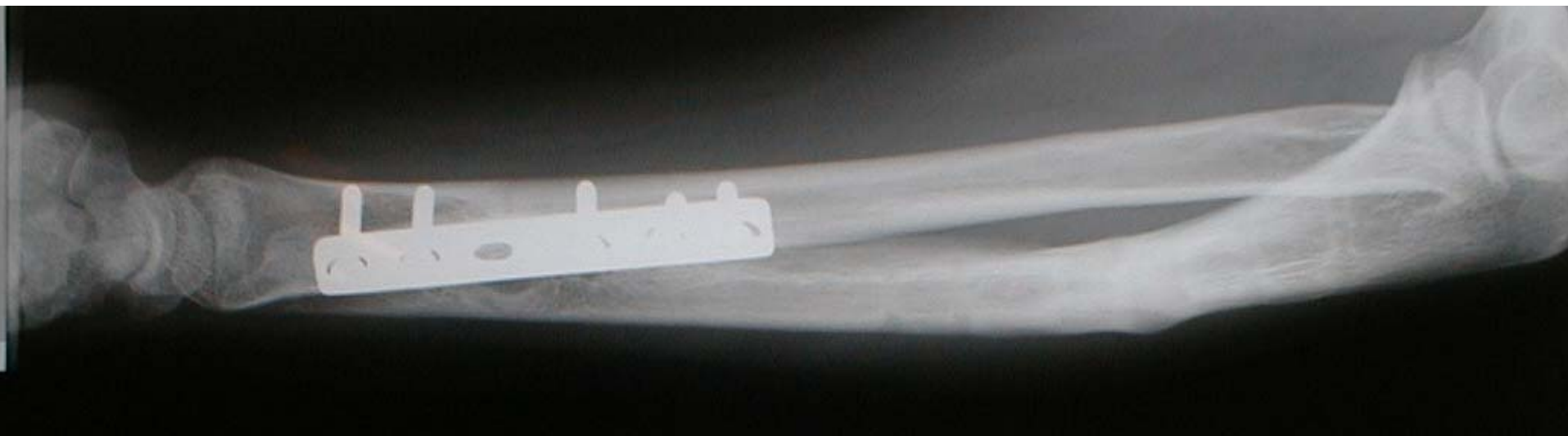


Fig.4E

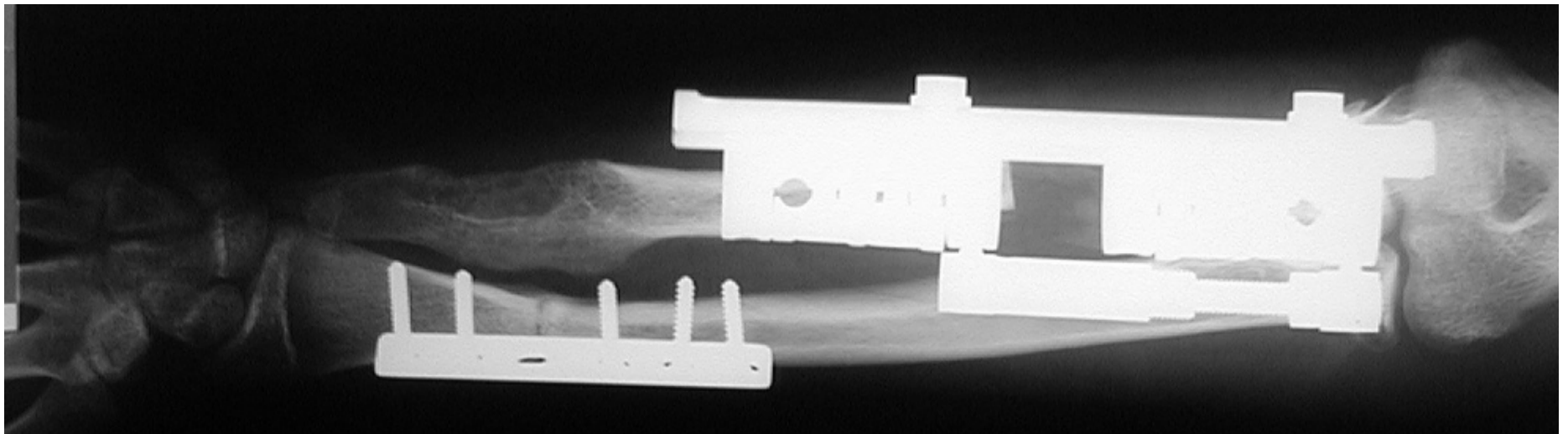
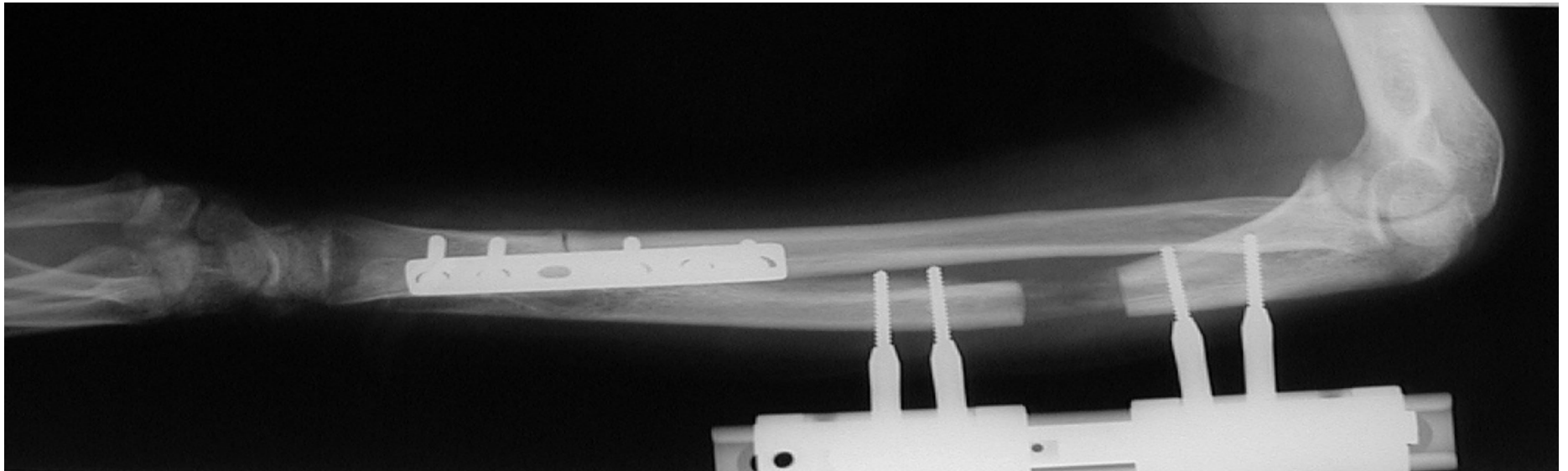


Fig.4D

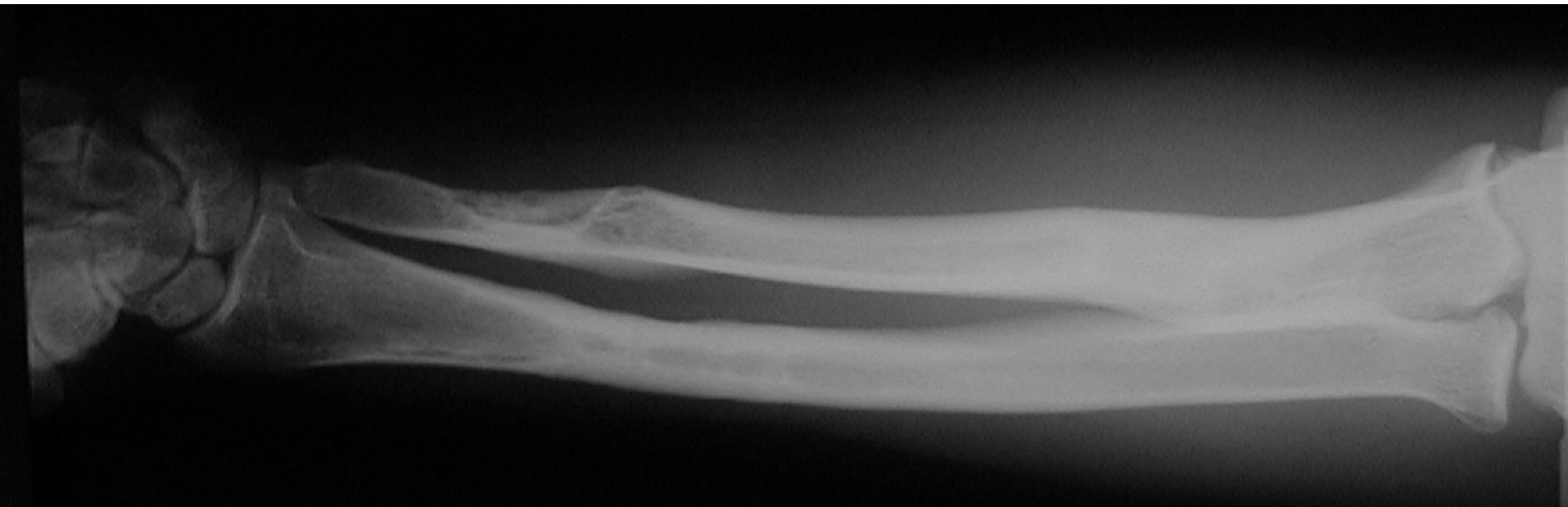
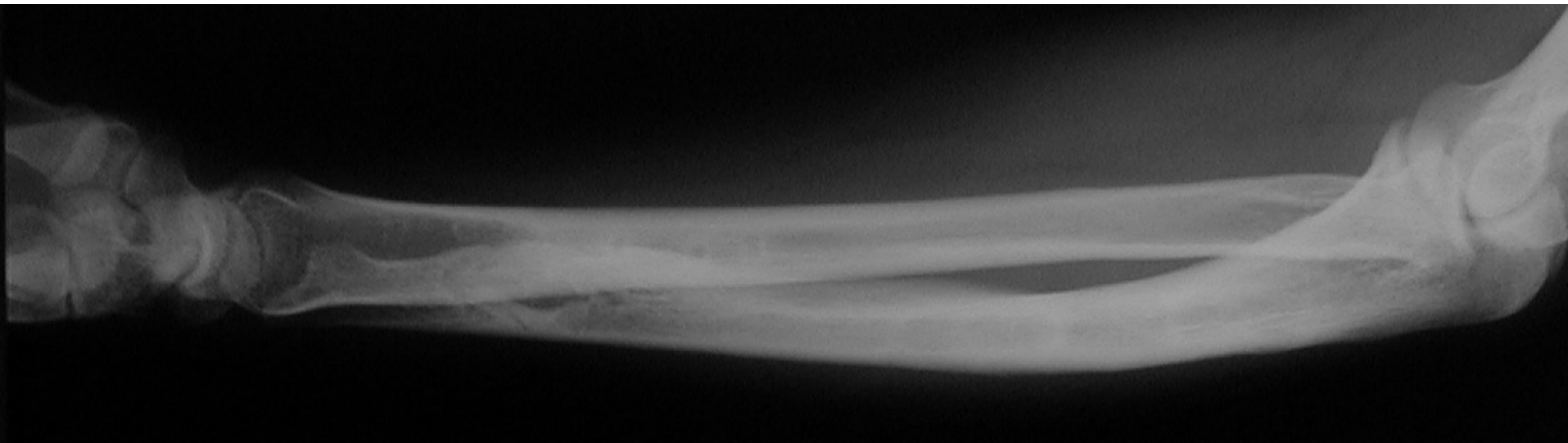


Fig.4G



Fig.4F

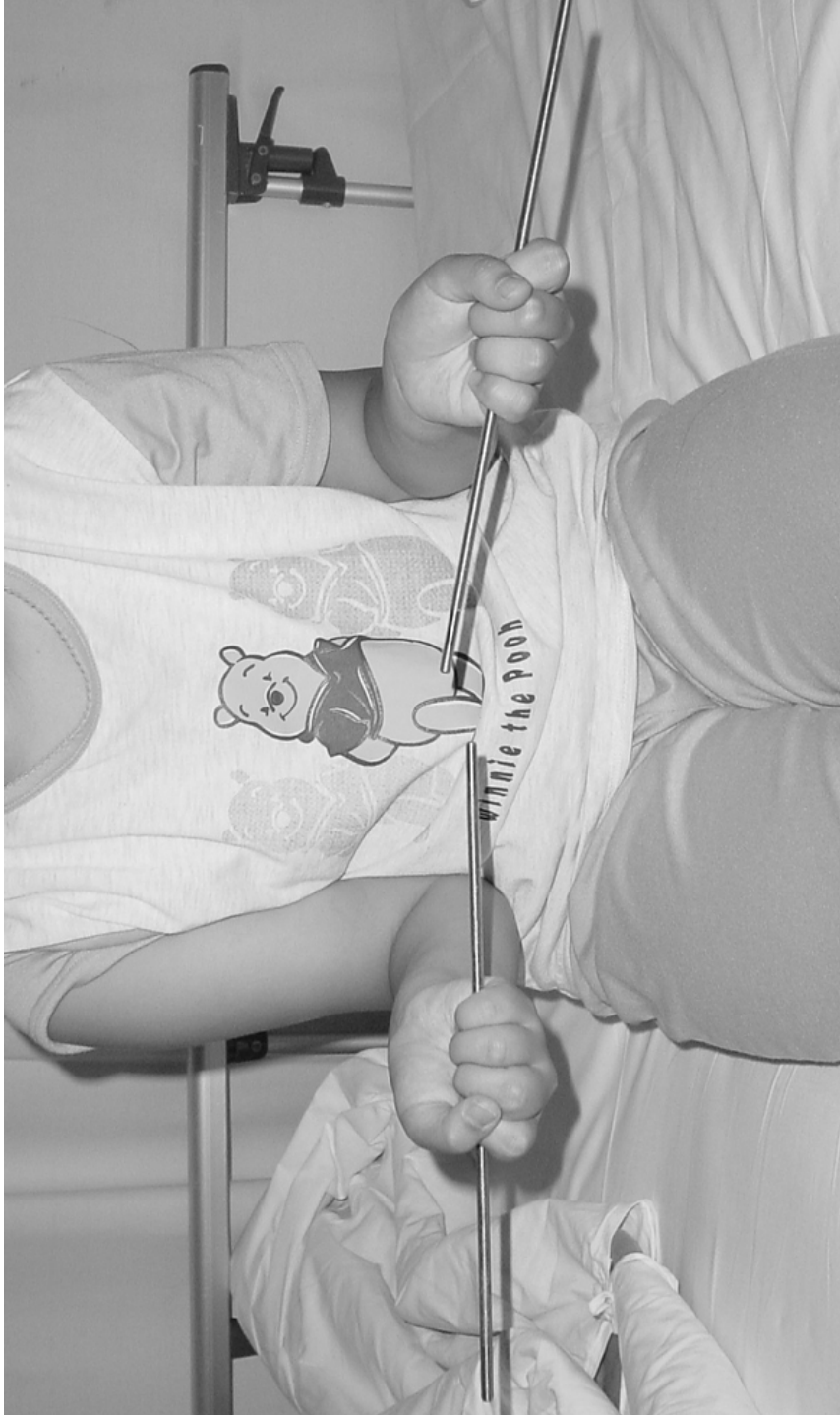


Fig.5A

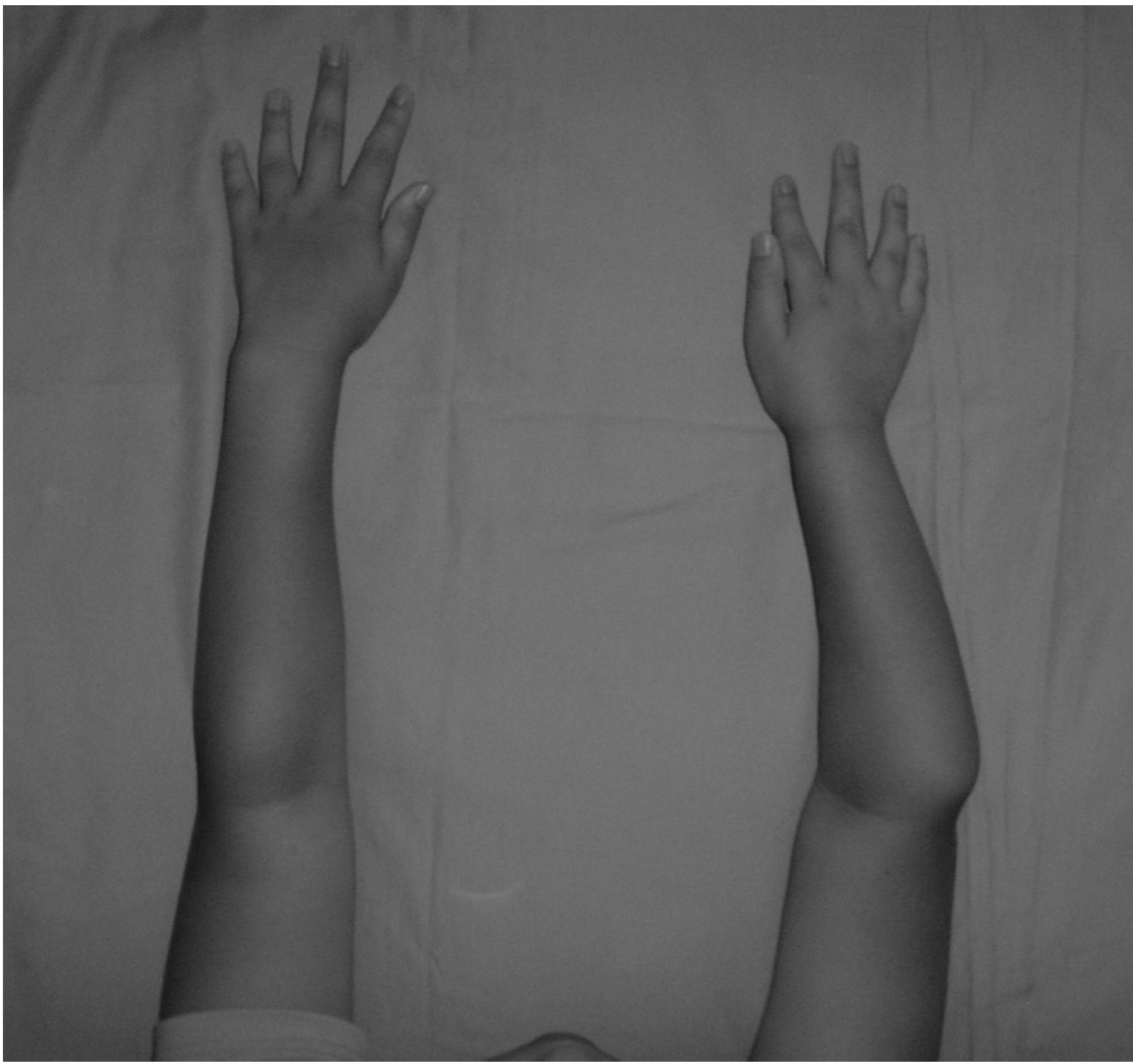


Fig.5B

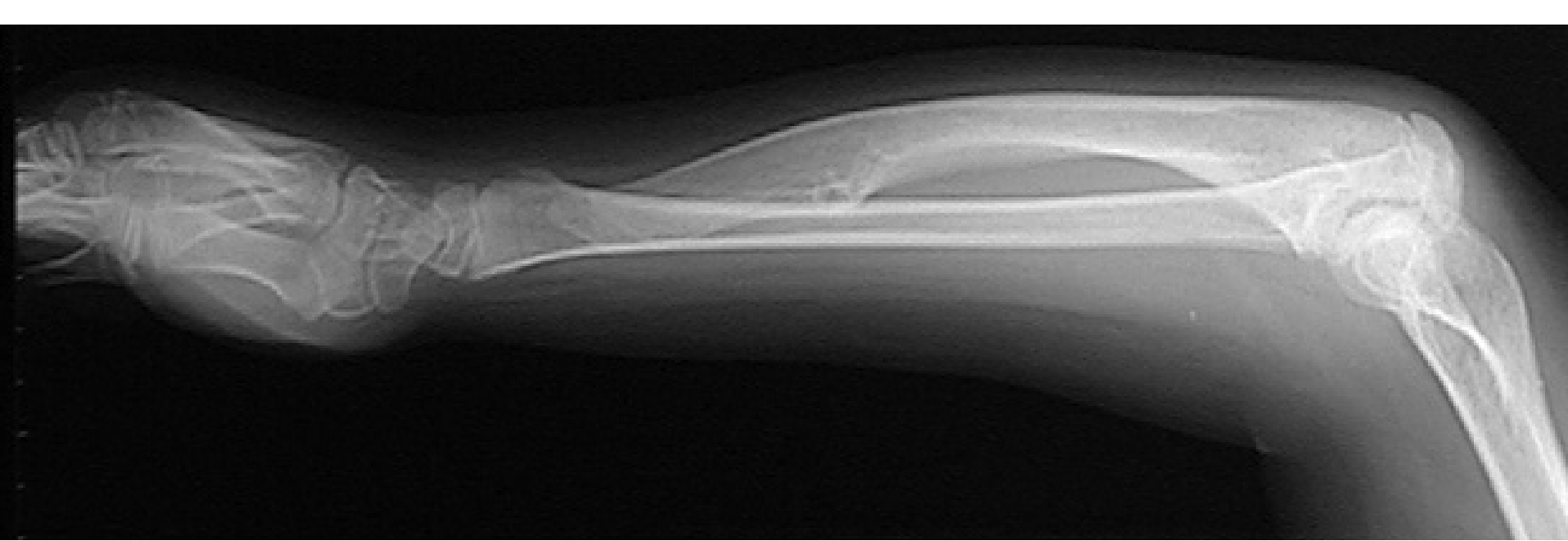


Fig.5C

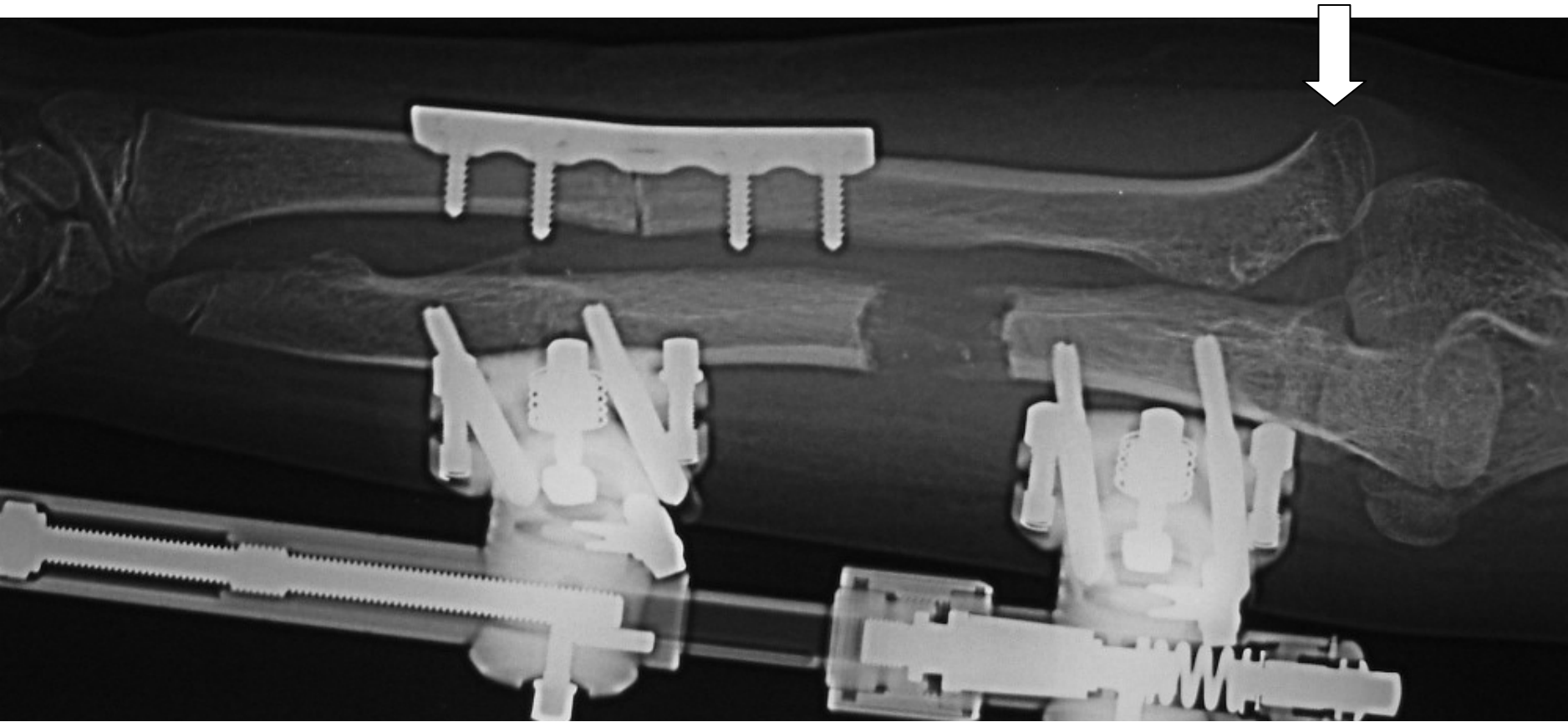


Fig.5D

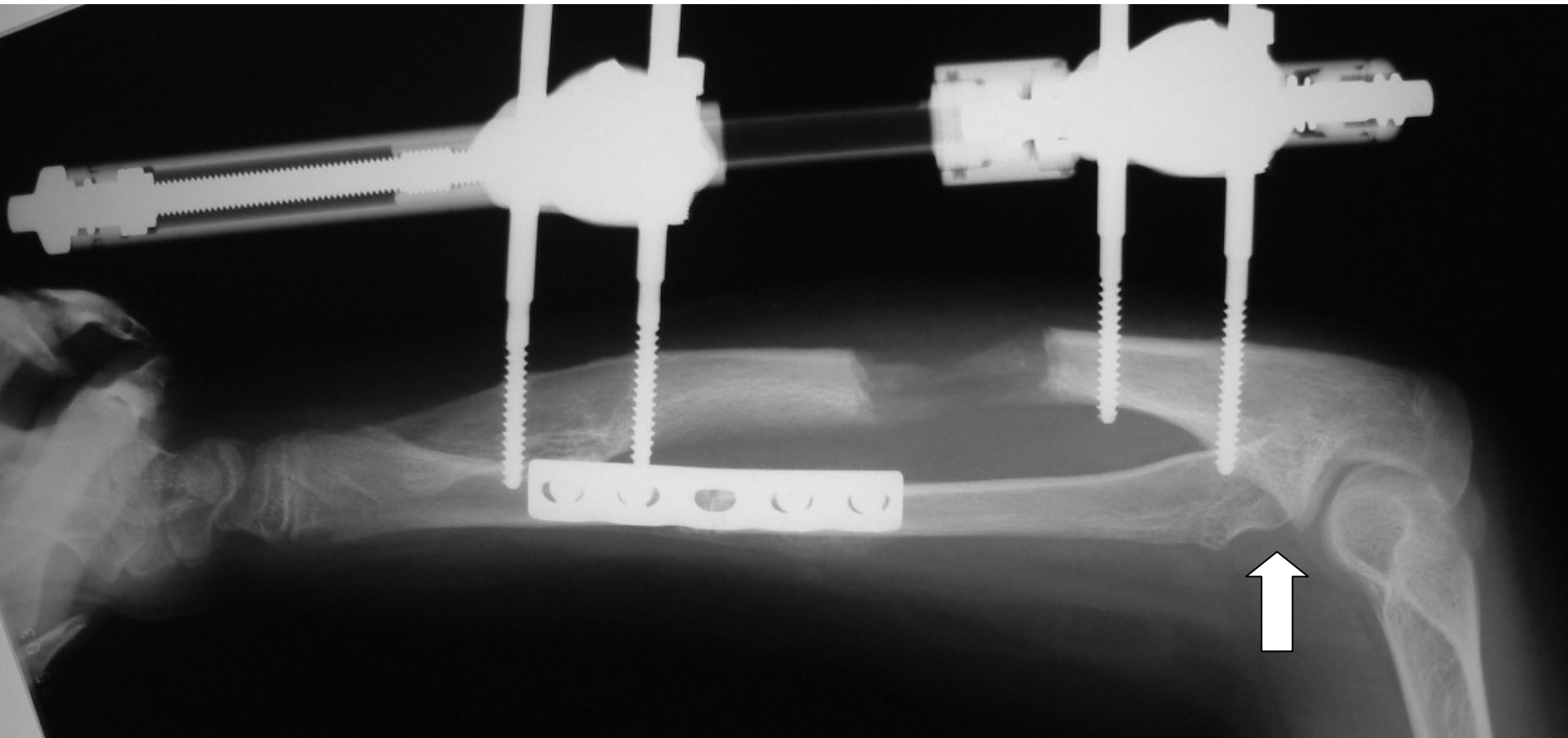


Fig.5E

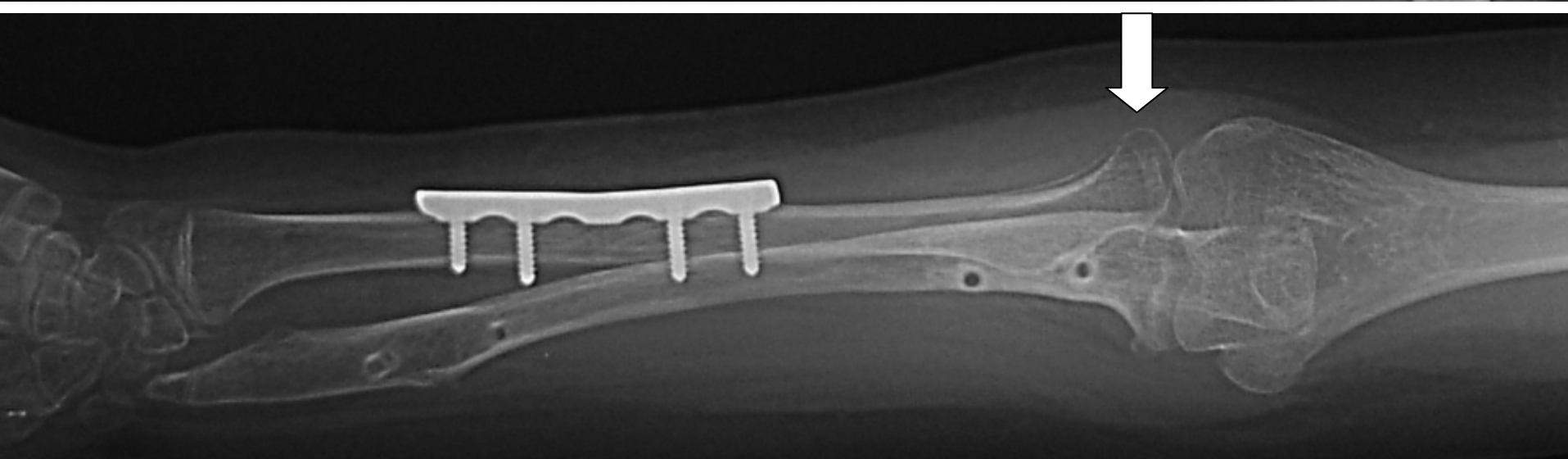
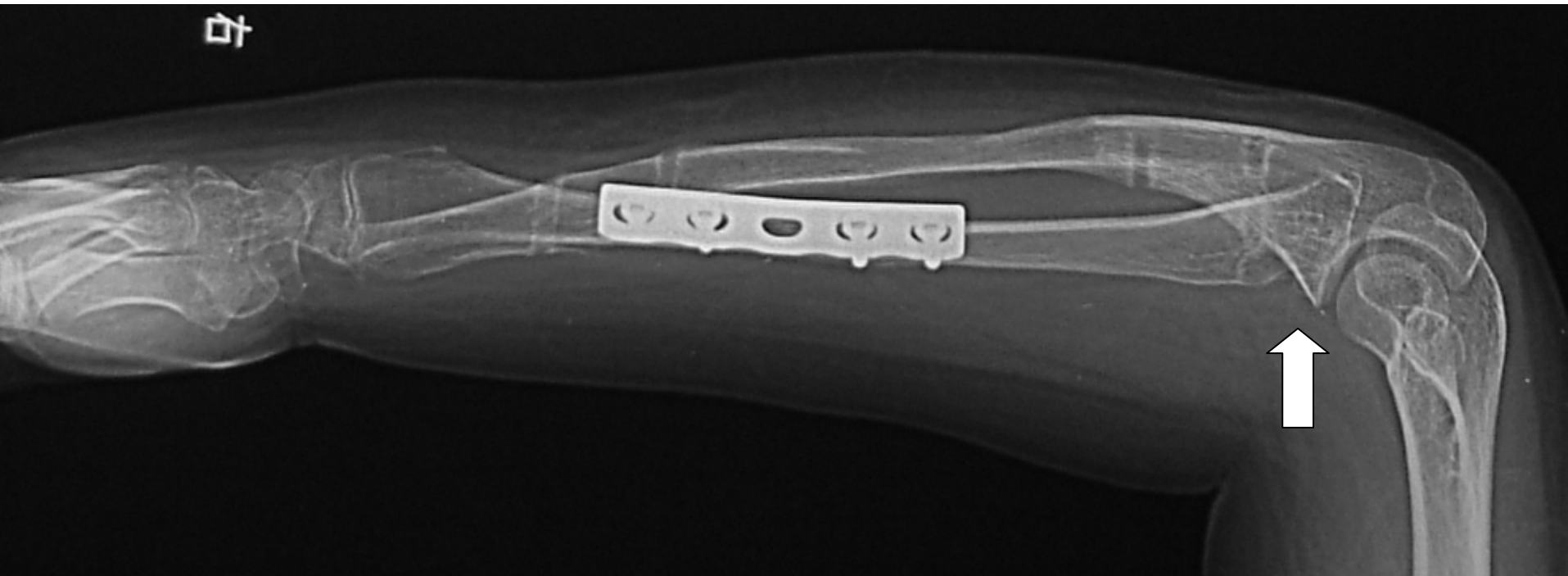


Fig.5F



Fig.5G

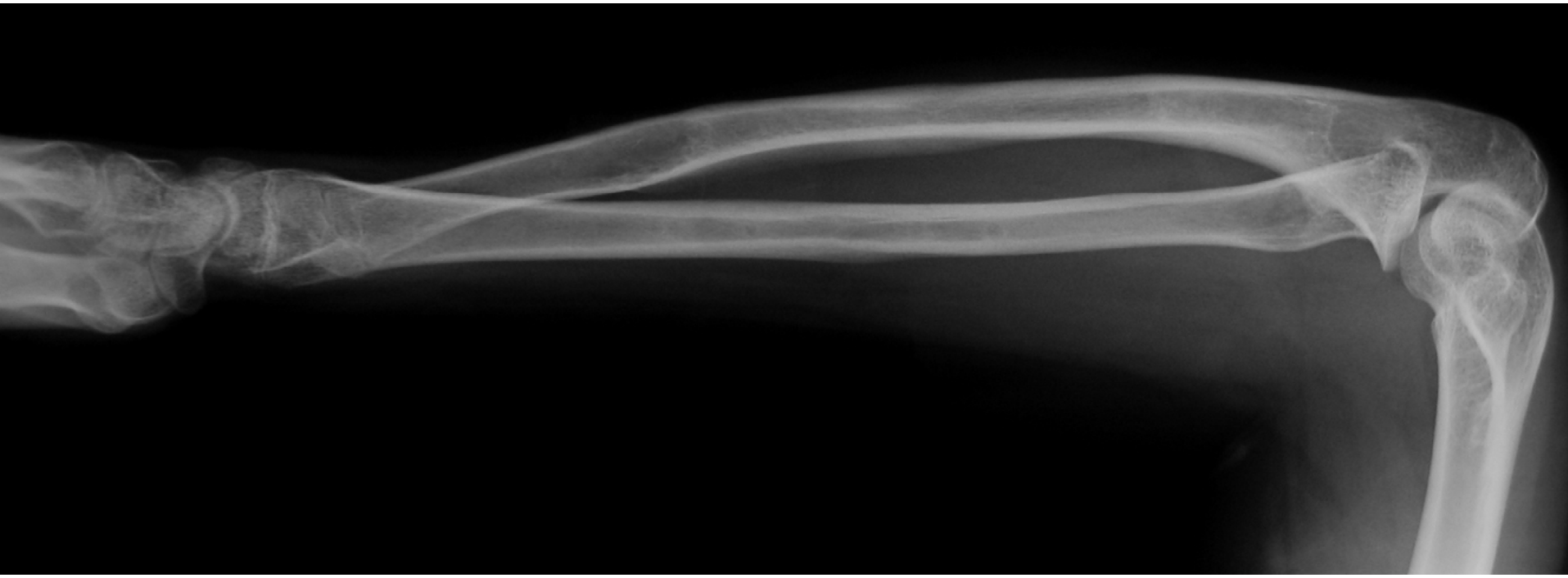


Fig.5H