filtration patterns of GAG were same in SED patients as in healthy persons. The value of serum sulfotransferase activity was normal in all SED cases. These discrepancies are now under consideration.

S - 5 骨系統疾患に対する脚延長術の導入

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[はじめに]

Achondroplasia症は四肢短縮型低身長症を呈する骨系統疾患の原型であり、全国に4000人以上、患者が存在すると推定される。知能は正常で生命の予後は良好、関節の機能障害も比較的少ないが、成人身長が120-130cmと低いため日常生活、社会生活に種々の制約を受ける（Fig. 1）。本質的な治療法のない現在、手術的に四肢を伸ばすことを希望して整形外科を受診する患者は少なくない。防衛大整形外科では achondroplasia症を初めとする四肢短縮型骨系統疾患に対し、DeBastianiらの方法にもとづき仮骨延長法による脚延長術を行なっているので、

Fig. 1. 15-year-old patient with achondroplasia. A prototype of short limb type skeletal dysplasia.

Fig. 2. Orthofix external fixator applied on ipsilateral femur and tibia

<table>
<thead>
<tr>
<th>Waiting</th>
<th>Distraction</th>
<th>Neutralisation</th>
<th>Dynamisation</th>
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<tbody>
<tr>
<td>←15days →</td>
<td>47days</td>
<td>45days</td>
<td>182days</td>
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Fig. 3. The schedule of bone lengthening by callus distraction
その臨床成績と問題点につき報告する。また、
手術方法や延長条件などを検討する目的で行った動物実験の結果についても併せて報告する。
[仮骨延長法（Callus distraction）の原理]

仮骨延長法とは、長管骨の骨幹部を手術的に
骨切りした部位に形成される仮骨を創外固定器
を用いてゆっくりと引き伸ばす方法であり、骨
移植を必要としない。一日1mmずつというゆっくり

Fig. 4. Radiographic time course of femoral lengthening
a) waiting period, b)–d) distraction period, e) neutralisation/dynamic load period,
f) after pin removal.
Fig. 5. X-ray finding of 18-year-old male with hypochondroplasia
Left femur and tibia were lengthened 7.5 cm each.

Fig. 6. Fourteen-year-old female with achondroplasia. Note improved alignment of left leg lengthened 13 cm.
Fig. 7-10. Experimental bone lengthening in rabbits
During postoperative waiting period, revascularization occurred around the osteotomy site (Fig. 7). Slow axial distraction (0.25 mm / 12 h) gave rise to elongation of immature callus (Fig. 8). As distraction advanced, a characteristic zone structure resembling growth plate was produced. Central radiolucent zone contained elongated fibrocartilage, while adjacent sclerotic zones consisted of fine cancellous bone (Fig. 9). Too fast distraction (1.0 mm / 12 h) caused disruption of the tissue. The callus formation was seriously disturbed (Fig. 10).
S·5 Clinical Experiences in Leg Lengthening by Callus Distraction

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and Y. SHIMOMURA

Sixty bony segments (29 femur and 31 tibia) in 21 achondroplastic patients (average age of 15 years; range 11 to 19 years) were lengthened by means of callus distraction. The average increase in limb length was 11.9 cm (range 8.5 - 16 cm).

In all cases, Orthofix unilateral lengtheners with four self-tapping screws were applied to the medial aspect of tibia and on the lateral aspect of femur. Subperiosteal transverse osteotomy was performed 1 cm below the second proximal screw with an electric power saw. After the postoperative waiting period for two or three weeks, distraction was commenced at the rate of 0.5 mm every 12 hours. Distraction was continued until active motion of the knee or the ankle joint was severely disturbed. After stopping distraction, there was a “neutralisation period” of 6 weeks on the average, during which full weight-bearing with the aid of crutches was encouraged. The locking screw of the lengthener was then loosened to give a dynamic axial loading for the next 6 weeks. The lengthener was removed after bony consolidation was confirmed by radiological and clinical examination.

The average overall treatment time was 182 days (range 110 to 260 days). In most cases, the range of motion of the knee and the ankle joint was considerably restricted during the distraction period, but completely restored after removal of the lengthener. Mild pin-track infection was noticed in 35% of the patients, most frequently around the proximal screws in the femoral component. There was no bony infection, no nerve complication and no vascular lesion. No case required a bone graft.

In conclusion, we believe that callotasis is a very safe method to use in lengthening the limbs of achondroplastic patients.