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Limb Lengthening for Short Stature: A 10-year Clinical Experience

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Abstract

Achondroplasia and hypochondroplasia limb lengthening has become better accepted over last thirty years. We studied our experience on all limb lengthening for short stature over the last ten years. We reviewed patient records, X-rays and Dual Energy X-ray Absorptiometry (DEXA) scans to evaluate the changes in body proportion after limb lengthening and the complications. Fourteen patients with 14 tibial, five femoral and two humeral lengthening procedures were included in this study. Average age at first lengthening procedure was 12.2 (9.6-14.4) years. The average height gained from tibial lengthening alone was 10.47 (2.8-16.3) cm. The average height gained from tibial and femoral lengthening was 19.4 (14.7-25.3) cm. The average humeral length gained was 11 (10.5-11.5) cm. All humeral and femoral lengthenings were performed with the Orthofix uniplanar fixator. In femoral lengthening, mild varus deformity occurred in all patients. Mild pin deformity occurred in all patients. Revisions were not required. More rigid pins are required for femoral lengthening. Tibial lengthenings were accomplished with three different types of fixators. Most complications were encountered in tibial lengthenings. Premature fusion occurred in three patients. One procurvatum and five valgus deformity occurred. Revision to multiplanar construct eliminated deformity. Body proportion has improved between upper and lower segments. Sitting to Standing height ratio decreased from 0.64 (0.59-0.74) to 0.58 (0.54-0.61). Armspan to Standing height ratio worsened from 0.92 before lengthening to 0.84 at follow up and did not have any significant functional effect. The ratios for an achondroplasia patient is 0.72 and 0.84 respectively. Callotasis BMD rises to 40% of original bone indicates premature fusion.

Key words Achondroplasia; Femur; Humeral; Lengthening; Tibia

Introduction

Limb lengthening for short stature patients has become less controversial as technical improvements had made such treatment safer, more reliable and with less complications.[1] The modifications of treatment methods and design of devices evolve with better understanding gained from clinical outcome studies and applied researches. Many types of external fixators had been developed since the Ilizarov revolutionary invention of his primitive circular fixator from bus parts in the 1950.[2] The basic principles of distraction osteogenesis Ilizarov discovered remained unchanged and still demanding further understanding. The methods of fixation, rate of distraction, assessment of bone formation (mineralisation) rate are still areas of research and clinical refinement. We review our experience over the last ten years to assess the outcome, complications and the body proportion changes after limb lengthening.

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Materials and Methods

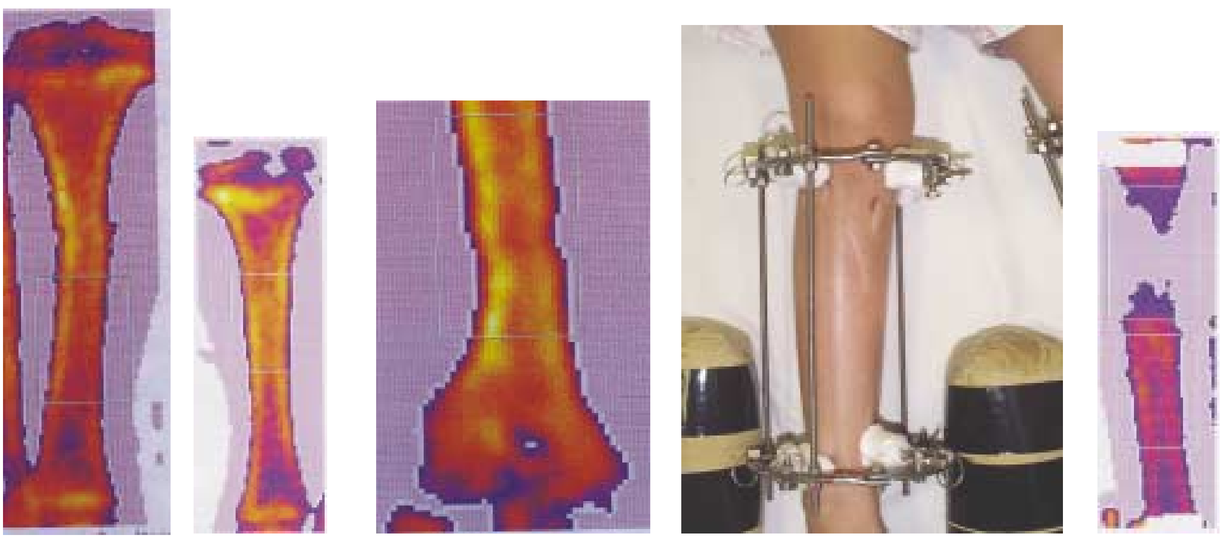
We reviewed all achondroplasia and hypochondroplasia limb lengthenings over the last 10 years. We studied clinical records, X-rays and Bone Mineral Density (BMD) changes as measured by Dual Energy X-ray Absorptiometry (DEXA) scans. We collected data on age, body weight, anthropometric measurements, fixation frame construct, rate of lengthening, the BMD changes during distraction and other interventional procedures. DEXA[3-6] scans had been used extensively for monitoring mineralisation of lengthened limb in this unit. The standard scans done include pre-operation on humerus as control, both tibiae at a marked site. The same sites were all scanned serially with reference to fixed distance from fixation pins to ensure consistency of the measured segment (Figure 1). The choice of fixation was selected by surgeon to be the best frame construct at the time according to the presence or not of deformity requiring correction or straight lengthening alone. Ilizarov hybrid circular fixators were used if deformity were present. The Orthofix uniplanar fixator were used for straight lengthening with no pre-existing axial deformity. It is used in all femoral and humeral lengthenings. These were applied according to DeBastiani method of limb lengthening by callus distraction – callotasis.[7]

Tibial Lengthening Operative Procedure

All patients underwent a standard sequence of treatment. After endotracheal intubation and general anaesthesia, a Foley catheter was passed and secure out of surgical field. Both operated upper or lower limbs were prepared and draped free to allow total access and X-ray screening. The limb axis and anatomical landmarks were marked on the limb surface. In tibia, these included patella, physeal lines, joint lines, dorsalis pedis and posterior tibial pulses. A syndesmosis screw was first placed across the ankle. A fibula osteotomy was performed by excision of approximately one cm segment. Fixation of proximal and distal segments were then performed. Osteotomies were made either by multiple drill holes conventional osteotomy or Ilizarov corticotomy. For humeral and femoral lengthening, Orthofix fixators were first applied in a standard manner. An osteotomy was performed at most proximal region of the bone segment.

Callotasis – Callus Distraction Lengthening

Post operation distraction was started 10 days after operation at one mm per day in four steps. The distraction rate was reduced when soft tissue tightness occurred, which responded temporary to stopping distraction for a few days. It indicated the end of lengthening.



a) Preoperative BMD scan of right tibia.

b) Preoperative BMD scan of left tibia.

c) Preoperative BMD scan of humerus as control.

d) Strict positioning of limb during lengthening ensures consistence of scan.

e) Measurement of bone segments according to fixed distance from fixation pin.

Figure 1 Scan technique & graphs.

Physiotherapy

All patients walked full weight bearing and use upper limbs fully by one to two weeks when they had recovered from initial wound pain and accustomed to the fixator. All patients had intensive range of motion exercises to all joints. Modified long arm splints and Knee Ankle Foot Orthosis (KAFO) built up on fixator were used as resting splintings to keep elbow and knee in extension and ankle in plantigrade during distraction. Frequent BMD study of the callotasis and the original host bone were done using DEXA scans weekly and two weekly during distraction and neutralisation respectively.

Termination of Lengthening

Distraction was stopped when there was increasing soft tissue tightness leading to joint motion range reduction not regained after a period of rest or progressive osteoporosis of the stress shielded original bone segments with BMD dropped to 20% of the starting level.

Neutralisation – Consolidation of Callotasis and Additional Treatments

During this consolidation period, the callotasis mineralisation as monitored by DEXA scan would progressively increase. Patients were encouraged to increase full weight bearing walking exercises. Dynamisation were instituted when continuous bone column were seen radiologically and DEXA scan showed no increase in mineralisation. This involved placement of elasticated spacer in the distraction device or progressive removal of fixation pins. Low intensity pulse ultrasound stimulation LIPUS and bone marrow injection was used when mineralisation failed to increase despite dynamisation.

Frame Removal

The fixator was removed when the BMD of Callotasis reached a plateau above 50-65% of original bone, the Callotasis was painfree on stress, and on X-ray showed a continuous well-formed bone column. After removal of the fixator, a well-moulded long arm orthosis was worn for upper limb. For lower limbs, a long leg dynacast was applied for six weeks followed by hinged KAFO for approximately six weeks or a well-moulded KAFO for twelve weeks.

Results

Fourteen patients were treated with one, two or three lengthenings. There were nine girls and four boys, the average age was 12.2 (9.6-14.4) years at the time of first lengthening procedure. All patients first had bilateral tibial lengthening, one patient had twice tibial lengthening (CMY). Five patients (COP, CMY, MHY, TWY, YSM) then had second stage femoral lengthening and two patients (COP, CMY) had third stage humeral lengthening at the present time. The average tibial lengthening including second lengthening was 9.57 (2.8-14.5) cm and 9.77 (2.8-14.5) cm; the Lengthening Index were 32 (19-58) and 30.3 (19.9-48.6) days/cm for right and left tibia respectively. The average femoral lengthening was 6.58 (4-9) cm and 6.74 (4.4-9) cm; the Lengthening Index were 35.2 (30.9-38.5) days/cm and 34.1 (30.9-35.7) days/cm for right and left femur respectively. The average lengthening for humerus were 11.25 (11-11.5) cm and 10.8 (10.1-11.5) cm; the Lengthening Index were 19.4 (18.7-20.1) days/cm and 20.2 (20.1-20.4) days /cm for right and left arm. The average gain in height from tibial lengthening, femoral and both were 10.47 (2.8-16.3), 6.74 (4.4-9) cm and 19.14 (14.7-25.3) cm respectively (Table 1).

Use of Fixators and Initial Deformity

All five bilateral femoral and two bilateral humeral lengthenings were done with Orthofix uniplanar fixators. Three pins per segment was used for femoral lengthening. Two pins per segment fixation were used for humeral lengthening. No complications were encountered in humeral lengthening. One frame revision was required for one of the femoral lengthening with no adverse effect. All the fixation pins deform at the end of femoral lengthening with mild varus deformity. No revision was required. The final limb alignment remained good with mechanical axis within one and a half cm of knee centre. For tibial lengthenings, two patients were treated with Orthofix monolateral fixator (OF). These were fixed with two or three 6 mm diameter rigid pins per segment. One patient was treated with the conventional all wires Ilizarov fixator (IO). Nine patients (10 lengthenings) were treated with Ilizarov Hybrid Advanced fixator (IHA). A combinations of transosseous tensioned wires and one or two parallel or crossed six-millimetre diameter half pins were used for each segment fixation (Table 2). Seven patients (NCY, LKY, MHY, LKK, KPH, YSM, CCKN) initially had one rigid 6/5 mm Orthofix pin fixation per segment (Stiffness category one S1P). Five of them developed valgus deformity. Two patients (LKK, CCKN) were revised to two crossed pin fixation with elimination of the deformity and recurrence. The valgus deformity in other three patients were mild and not requiring correction. Four patients

Table 1 Height gain from tibial and femoral lengthening

	Height gain from tibia (cm)	Height gain from femur (cm)	Total gain leg length (cm)	Height gain from tibia & femur	Humeral length gain
CCKN	12.3		12.3		
CKL	10.5		10.5		
CMY	16.3	9	25.3	25.3	
COP	14	5.7	19.7	19.7	11.5
KPH	9.3		9.3		10.5
LKK	12.2		12.2		
LKY	10		10		
MHY	10.1	6.5	16.6	16.6	
NCY	8.7		8.7		
SCK	9.1		9.1		
TWY	11.3	8.1	19.4	19.4	
WCY	2.8		2.8		
YSM	10.3	4.4	14.7	14.7	
YTK	9.7		9.7		
Average	**10.47**	**6.74**	**12.88**	**19.14**	**11**

Table 2 Axial deformity and frame construct relationships

Name	Ex fix type	Construct	Rigidity rating	No. of callotasis	Complications
KPH	IHA	2w1p+2w1p	R&L S1P1p	1	Additional pin for procurvatum
MHY	IHA	R 2w1p+2w1p, L3w1p+2w1p	R&L S1P1p	1	Bilateral Valgus deformity, premature fusion, osteotomy crack up pin, PETA
YSM	IHA	R&L (2w1p+2xp+2w1p)	R&L S1P1p	2	No valgus deformity
NCY	IHA	R&L (2w1p+2w1p)	R&L S1P1p	1	Osteopenia
LKY	IHA	R&L (2w1p+2w1p)	R&L S1P1p	1	Bilateral valgus deformity
LKK	IHA	R&L (2w1p+2xp+2w1p) revised to R&L (2W2XP+2XP+2W1P)	R&L S1P2xp rev to S2XP2xp	2	Valgus deformity, frame revision to 2 XP, PETA
CCKN	IHA	R&L (2w1p+2xp+2w1p) revised to R&L (2W2XP+2XP+2W1P)	R&L S1P2xp rev to S2XP2xp	2	Premature fusion, callotasis osteotomy, PETA
TWY	IHA	R&L (2w2pp+2w2pp)	R&L S2PP2pp	1	Valgus deformity, BMI
CMY1	IHA	R&L (2w2xp+2w2xp)	R&L S2XP2xp	1	BMI
CMY2	IHA	R (1W2Xp+1w 2AP) L (1w2pp+1w2pp)	R (S2XP2pp) L (S2PP2pp)	1	Valgus deformity on Left
COP	IO	ALL WIRES	R&L S01X	2	No valgus deformity
YTK	OF	R&L (2pp+2pp)	R&L S2PP2pp	1	Valgus deformity, post frame off fracture
SCK	OF	R&L (3pp+3pp)	R&L S3PP3pp	1	Valgus deformity

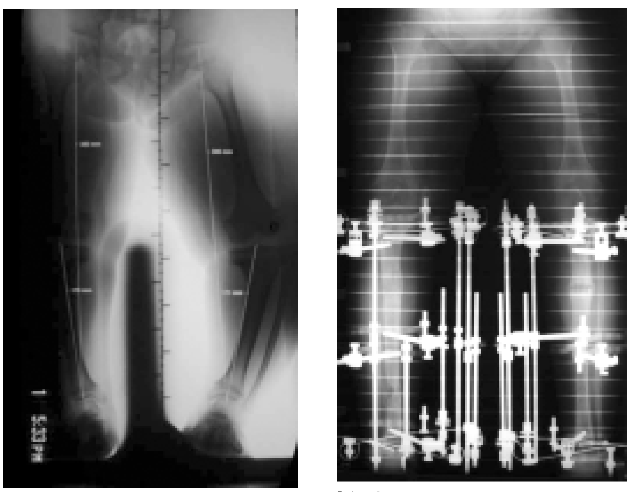
(CMY1, CMY2, TWY, YTK) had fixations consisting of two rigid pins per segment (Stiffness category two S2PP or 2XP-PP represents parallel pin and XP represent crossed pin). All limbs fixed with parallel pins developed valgus deformity. None of the limbs fixed with crossed pins developed valgus deformity. One patient (SCK) had fixation with three parallel rigid pins (S3PP) per segment of fixation. She had also developed valgus deformity.

Tibial Lengthening and Correction of Knee and Ankle Varus Joint Orientation

Varus knees and ankles are common deformities in achondroplasia patients. The over growth of the fibula caused lateral collateral laxities of the knee and ankle. During the process of lengthening we had observed the fibula callotasis tend to consolidate after approximately 8 cm of lengthening. Continued tibial lengthening with this fused fibula callotasis caused the fibula head to migrate distally and tightens the lateral collateral ligament of the knee. The joint orientation then become horizontalised (Figure 2).

Tibial Lengthening Complications

Problems of pin tract infection happened to all patients. These responded to a short course of appropriate antibiotics and increased frequency of pin tract cleansing. Obstacles as defined by Paley[8] included one half pin required removal due to recurrent infection.



a) Antero-posterior X-ray showing varus knee and ankle.

b) Antero-posterior X-ray showing correction with distraction note distal descend of fibula.

Figure 2 Normalisation of joint orientation.

Tibial Fixation Revisions

Four patients required revision of fixation all at the proximal tibial segment. One patient (MHY) had corticotomy cracked proximally towards one of the fixation wire. This was revised on the third day after the check by X-ray. There was delay of distraction due to presence of pain. MHY developed two episodes of premature fibular fusion requiring repeat callus osteotomy. One patient (KPH) required placement of an additional half pin to control procurvatum deformity during lengthening. Two patients (LKK, CCKN) had revision for valgus deformity.

Tibial Additional Procedures

Two patients (TWY, NCY) had ultrasonic therapy and three patients (TWY, KPH, CMY) had bone marrow injections to callotasis site during the neutral fixation period when there appeared to have no progress of mineralisation. Three patients (YSM, LKK, CCKN) required percutaneous Tendo Achilles tendon elongation for loss of ankle dorsiflexion. One patient (YTK) suffered from post frame removal fracture. This patient developed extensive dermatitis towards the end of the treatment related to pin sites. Walking exercises were reduced due to these pin problems. Fixator was removed when the bone was osteoporotic. YKT developed axial deformity within a long leg cast. It healed with an acceptable alignment.

There were no fracture, vascular or neurological complications during distraction and neutralisaton period.

Tibial and Fibula Premature Callotasis Fusion

Two patients (MHY, CMY1) had premature fusion of the fibula callotasis requiring repeat osteotomy. One patient (CCKN) had premature fusion of tibial callotasis requiring callotasis osteotomy. The callotasis BMD for this patient rose sharply to 40% of the original bone associated with deformity of the distraction pin (Figure 3). The S1P fixation construct was also revised to S2XP for valgus deformity.

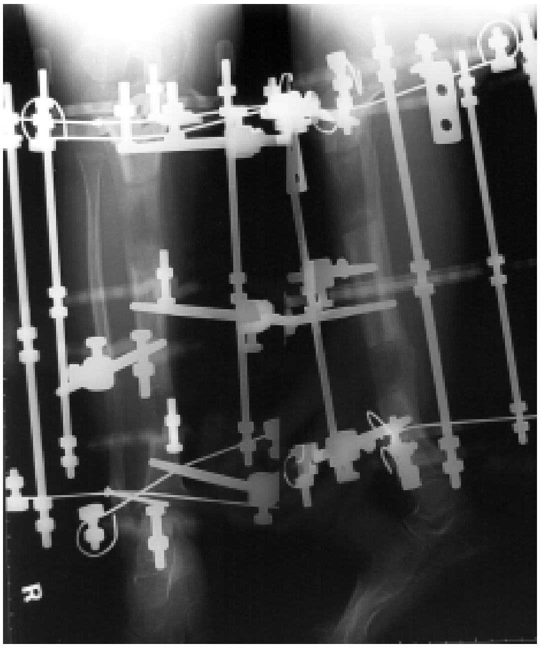
All patients were satisfied with the result of lengthening including the patient who had fracture.

Scar Release

All patient developed some pitting and tethering of the pin and wire tract scars. These were all released at the time of frame removal and formed an important part of the treatment under general anaesthesia.

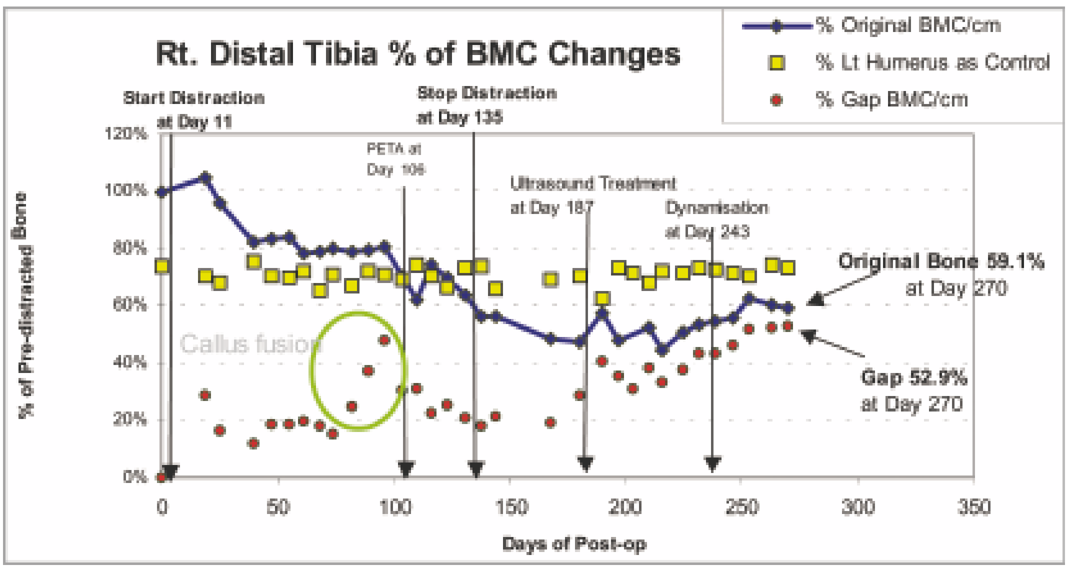
Changes of Body Proportion (Table 3)

The body proportion visually changes after patient had their tibial and femoral lengthenings (Figure 4). This can



a) AP & Lateral X-rays showing premature fusion of callotasis, note wire and pin deformity.

b) AP & Lateral X-rays showing rupture of callotasis with continued lengthening.

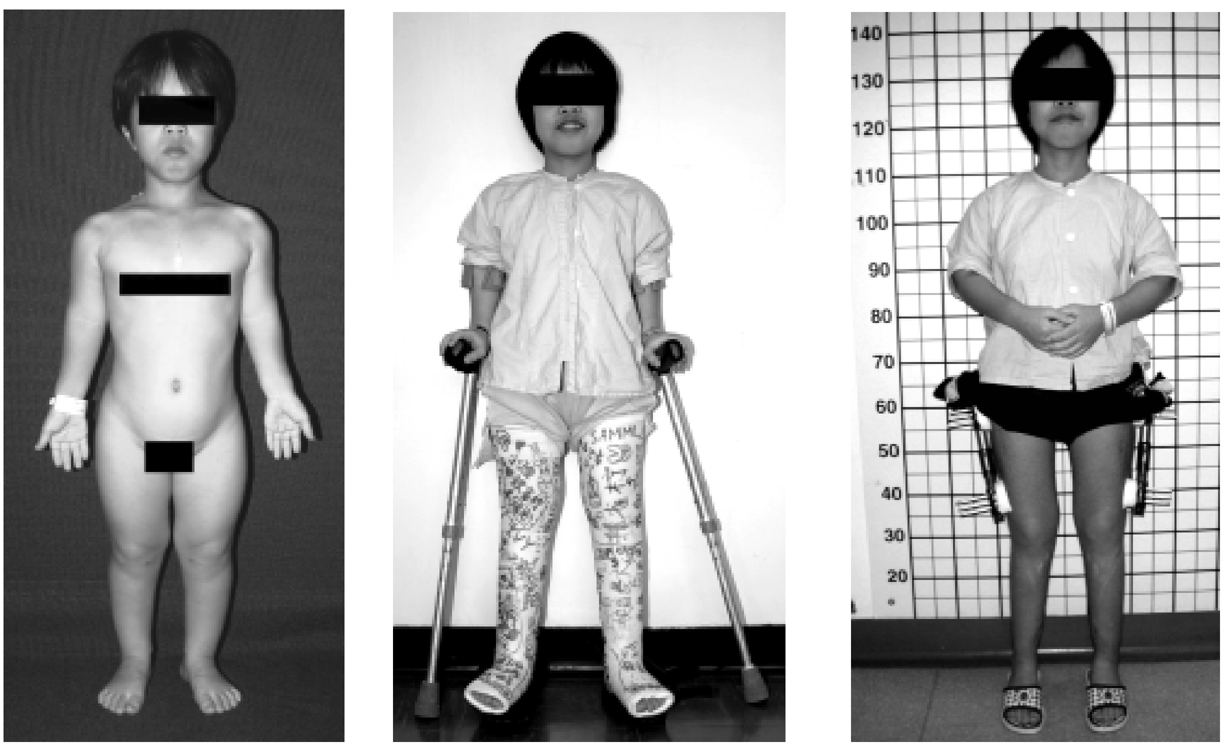


c) Sharp rise of callotasis BMD to 40% corresponds to fusion, sharp drop after rupture.

Figure 3 Callotasis BMD changes during lengthening.

Table 3 Body proportion changes with height gain

Standing ht Pre OP (cm)	Standing ht Final FU (cm)	Armspan Pre OP (cm)	Armspan Final FU (cm)	Seating ht Pre OP (cm)	Seating ht Final FU (cm)	Armspan/ Standing ht Ratio Pre LL	Armspan/ Standing ht Ratio Post LL	Sitting ht/ Standing ht Ratio Pre LL	Sitting ht/ Standing ht Ratio Post LL
CCKN 109	121	105	108	70	70	0.96	0.89	0.64	0.58
CKL 106	129	100	113	63.5	74	0.94	0.88	0.60	0.57
CMY 104	141	94.8	127	76.5	79	0.91	0.90	0.74	0.56
COP 96	130.5	80	112	60	76	0.83	0.86	0.63	0.58
KPH 116	128.5	109	110	68	70	0.94	0.86	0.59	0.54
LKK 107	127	100	101	70	76	0.93	0.80	0.65	0.60
LKY 106	119	98	102	70	72	0.92	0.86	0.66	0.61
MHY 105	126	97	104	68.5	75	0.92	0.83	0.65	0.60
NCY 101	119	90	102	64	73	0.89	0.86	0.63	0.61
SCK 130	142	129	132	78	80	0.99	0.93	0.60	0.56
TWY 106	137	98	110	70	77	0.92	0.80	0.66	0.56
WCY 124	132	119	125	80	80	0.96	0.95	0.65	0.61
YSM 125	137.5	114	114	80	81	0.91	0.83	0.64	0.59
YTK 109	132	90	116	63	76	0.83	0.88	0.58	0.58



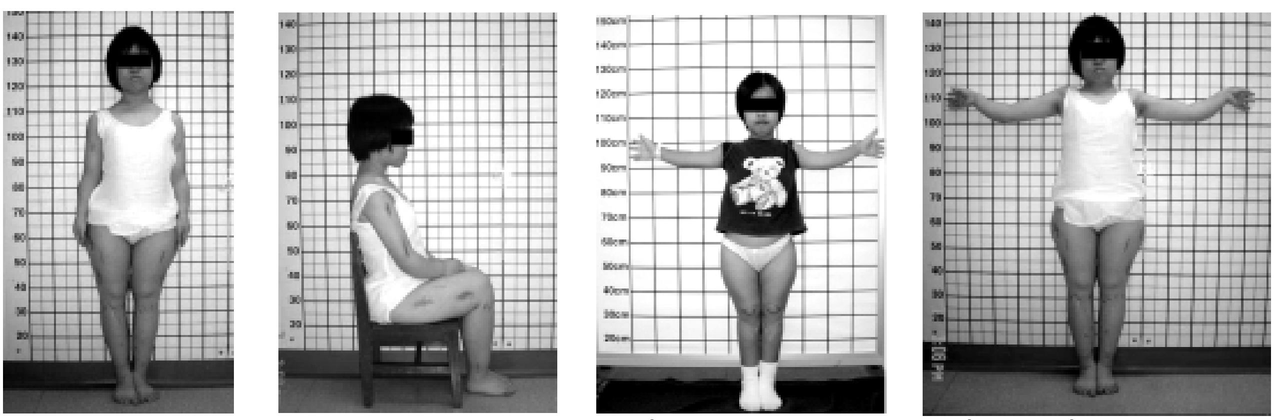
a) Before limb lengthening. b) After tibial lengthening. c) After tibial and femoral lengthening.

Figure 4 Achondroplasia patient body proportion changes from left to right.

be objectively measured by the sitting and standing height ratio. The arm span to standing height ratio worsened but it was mostly not apparent as the lower limb lengthening occurred below the hip and knee. The arms remained at the hip level on standing. The Sitting and Standing height ratio changed from an average of 0.64 (0.58-0.73) to 0.58 (0.54-0.61). The Armspan, Sitting and Standing height ratio(AS/SH) changed from an average of 0.92 (0.83-0.99) to 0.86 (0.83-0.95). These ratios were best restored close to normal when all three-tibial, femoral and humeral-lengthenings had been done (Figures 5a & 5b). The effect of Armspan to Standing height ratio however is not very apparent clinically comparing patients before (AS/SH 0.8) and after humeral lengthening (AS/SH 0.9) (Figures 5c & 5d). It makes a great difference to patient after humeral lengthening as patient can purchasing normal clothing without need to modify sleeve for the short arms.

Discussion

Bone lengthening consists of fixation of segments with an external fixation device, which allows gradual controlled separation of the fixed segments. An osteotomy is made between these segments. Limb lengthening dated back to 1920 when Codilla reported an one-stage technique. There were some good results and many failures and complications. Anderson[9] in 1952 reported a method of leg lengthening which resembled the modern method. The author described a procedure included first elongation of tendo calcaneus, second a long oblique or step cut osteotomy and third fixation by an Abbot type distraction frame with two fixation pins for each segment. Lengthening was achieved by slow screw distraction at a rate of one eighth of an inch (3.3 mm) per day. Wagner[10] in 1978 reported lengthening using a uni-planer fixator with a diaphyseal osteotomy. The treatment consisted of latency of five to seven days; distraction of 1 mm per day in single distraction; plate fixation at end of lengthening. Bone graft was frequently required. Fracture after plate removal had been a major complication. DeBastiani et al reported a similar treatment method with a metaphyseal osteotomy instead of a diaphyseal osteotomy in 1987. The distraction rate of one mm per day in four steps. They coined the term callotasis-callus distraction in limb lengthening. Ilizarov[11] from Russia invented the circular fixator for the treatment of limb deformities and shortening in 1950s. His monumental discovery of distraction osteogenesis capacity of bone revolutionised the limb lengthening treatment. Ilizarov used thin transosseous tensioned wires mounted on rings held by metal rods. He postulated six prerequisites that remained like principles of bone lengthening: 1) Preservation of periosteum, endosteum and marrow circulation by corticortomy; 2) Stable external fixation from tensioned wires; 3) A rest period before distraction allows the formation of fracture callus; 4) Small high frequency distraction stimulate direct bone growth; 5) The regenerate remodels under the influence of the tensile force; 6) Physiological limb use stimulates callus formation and ossification of the regenerate. The Ilizarov technique became famous in 1967



a) Hand position at neutral after humeral lengthening.

b) Sitting height, note limb ratio.

c) Before humeral lengthening.

d) After tibial, femoral and humeral lengthening.

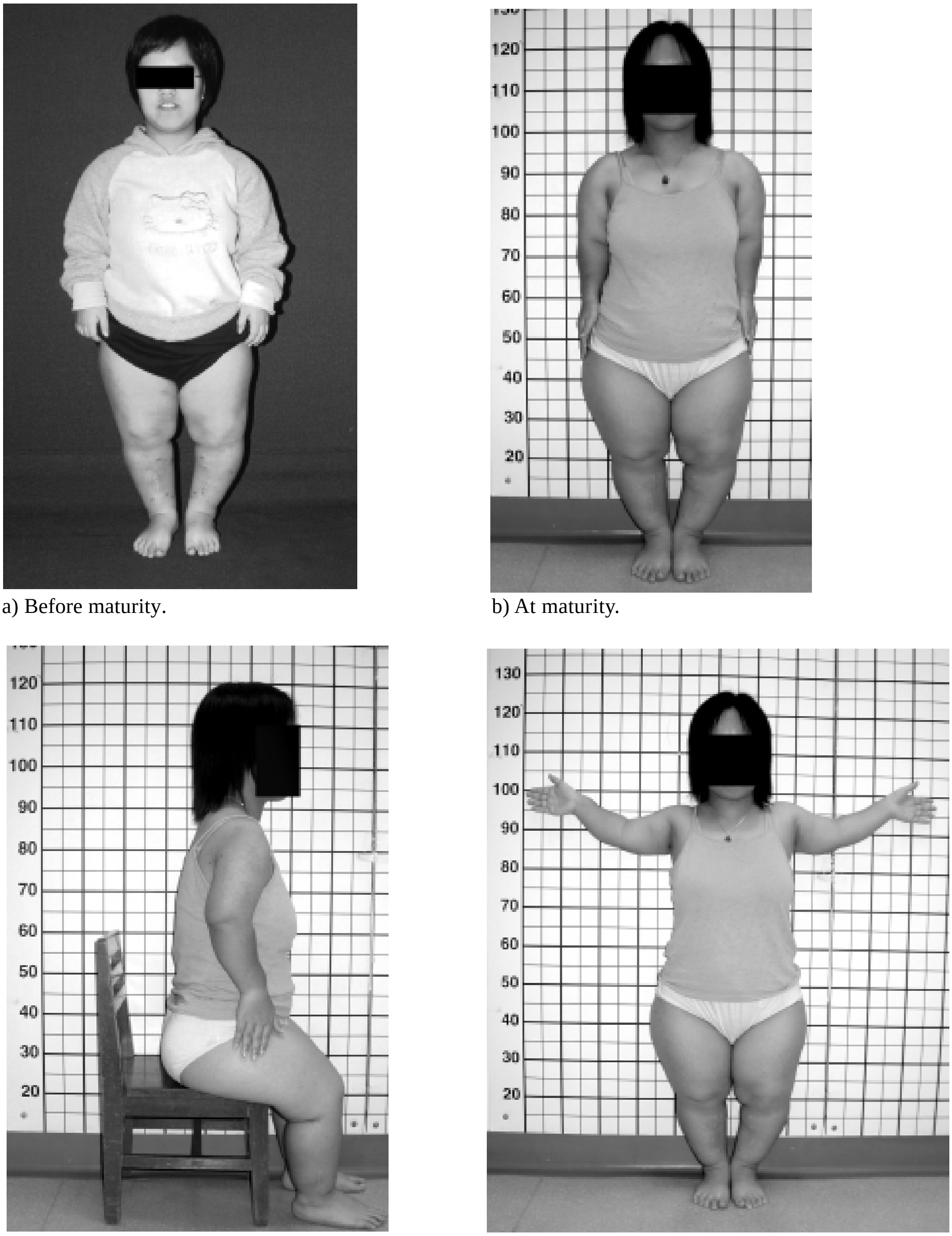
Figure 5 Armspan proportion in Achondroplasia patients after tibial and femoral lengthening.

when the famed Soviet high jumper Valery Brumel had been successfully treated for a tibial nonunion from compound fracture resulting from motor cycle accident. The Ilizarov method was subsequently introduced in Italy in 1981 and in the United States in 1987. This basic method had been used in all of our patients. The Ilizarov method was introduced to us by Dr D Bell who treated our first case by the conventional all wire transosseous tensioned wire fixation. The fixation included whole tibia and the foot with bifocal lengthening. The result of this patient (COP) had been excellent. However the fixation was complex, bulky in order to be stable[12,13] and uncomfortable to patient. Subsequent three cases were treated with uniplanar Orthofix fixator by DeBastiani callotasis method. The uniplanar fixator were chosen as there were no deformity. It was simple to apply and more comfortable as it does not immobilise the ankle joint. It only allowed monofocal lengthening. The unilateral fixators now had other improvements that would allow some rotation of the pin clamp on the sliding rod unit. This allows more freedom of bone fixation but it loses stability. In external fixation versatility and stability contradicts each other. In this study the rigid monolateral fixation pins were observed to deform in all lengthenings including humeral, femoral and tibial fixatons. Varus in femur and humerus and valgus in tibia deformities occurred. More rigid pins are therefore required. In tibial lengthening pin deformity may contribute to premature fusion. The later tibial lengthening cases were all treated with Ilizarov hybrid fixators. Some patients also presented with deformities. The Ilizarov and hybrid fixators are known as a multiplanar fixators that allow deformity correction and lengthening. The introduction of rigid pins greatly increase rigidity and comfort.[14-16] We had noticed the one pin or two parallel pins per segment fixation lead to valgus deformity during lengthening. The revision to a two-crossed pins per segment fixation prevented further valgus deformity.

It had been observed that the patients (MHY, CCKN, CMY) with a higher initial BMD had a shorter lengthening index and a higher chance of premature fusion. These had been termed "quick bone former". Premature fusion was observed when there were subtle pin deformity and a sudden rise of BMD in the callotasis. A static callotasis gap and the sudden rise in callotasis BMD to 40% of original bone signify premature fusion. This generally occurred when soft tissue tension increased with lengthening, the range of motion of the ankle dorsiflexion had been reducing. The distraction rate was reduced or stopped for the soft tissue to recover with intensive physiotherapy. This slowed distraction coupled with the quick bone formers will lead to premature consolidation. Proactive decision should therefore be made when range of motion was reducing. This involves elongation of the causative large muscle in the leg. The Tendo Achilles tendon can be elongated by a percutaneous technique. Distraction can be resumed at normal rate. Callotasis osteotomy would have to be performed if fusion was established and further lengthening was intended. We also learnt from one case (CCKN) that callus osteotomy healed slowly if performed at the mid segment of the regenerated bone. It may be better to create the osteotomy at the proximal end of the regenerate where the blood supply is better.

Lengthening Index is a measure of rate of healing. The decision on when to remove the frame has been same for all patients in this series. The increased frame rigidity seemed to be related to clinical reduction of rate of mineralisation of the callotasis and increased the osteoporosis of the stress-shielded bone. The bone mineral density of two patients (TWY, NCY) had dropped rapidly during distraction in the stress-shielded bone segments. TWY was believed to have too rigid fixation and too rapid a distraction rate (1.5 mm per day) at the beginning of the distraction when there was concern about premature fusion. She subsequently had progressive removal of fixation pin to dynamise the fixator with resulting increase in mineralisation. NCY had low body weight and low baseline Bone mineral density. This was a reflection of the nutritional and activity status of the patient. He had rapid drop in BMD of the stress-shielded bone. This was believed to be related to too rigid fixation with a two wires and one pin construct for this patient. The BMD recovered rapidly after removal of the fixator. Two wires and one pin per segment fixation seemed to be the best combination in terms of elasticity. Two wires two cross pins per segment fixation at maximal intersection angles seemed best for stability and rigidity of fixation. Early dynamisation is therefore advisable to maximise the rate of mineralisation particularly during neutral fixation when mineralisation reached a plateau. DEXA scan had allowed a close monitoring of this quantitative change in ossification of the regenerated bone and determine the timing of dynamisation and timely frame removal. We had not seen any difference in healing rate of regenerated bone between different types of osteotomies and corticotomy. The percutaneous corticotomy as advocated by Ilizarov is technically difficult particularly for second lengthening. We now use a Gigli saw osteotomy through two small incisions.

Achondroplasia patients has trunk length same as normal person.[17,18] The disproportion become most pronounced at adult hood. Therefore the present method of comparing Sitting height, Armspan and Standing height before and after lengthening was not entirely appropriate as the pre-lengthening ratio in immature patient is certainly not as abnormal as it is in achondroplastic adult. The Armspan and Sitting to Standing height ratio in an adult achondroplasia patient was 0.84 and 0.72 respectively Figure 6 (HCH). A ratio of close to 1 for Armspan/Standing height and 0.5 for Sitting/Standing height would be most balanced



a) Before maturity.

b) At maturity.

c) Sitting height, note short thigh length.

d) Armspan at maturity.

Figure 6 Achondroplasia patient body proportion.

proportion for Chinese.[19,20]

We had learnt from the treatment of every case. Limb lengthening is a complex treatment spanned over a long period of time. The ideal fixation is still to be investigated. This will have to take patient factors, frame construct into consideration. The rigidity should ideally be variable from a rigid beginning to an elastic end. The occurrence of pin deformity and premature fusion indicates that the current used pins may not be rigid enough to maintain distraction when the callotasis mineralisation increases to that level when fusion occurs. The use of a close monitoring system like the DEXA scan is indispensable. Its application will be exploited further. The limb lengthening method has also developed into lengthening on nail. This is intended to shorten fixator time. It does limit fixation stability and violates the intramedullary bone and circulation. More complications are expected from such method. Intramedullary lengthener[21] is also being developed where small wounds were used to place an interlocking intramedullary distraction rod. The osteotomy is performed with intramedullary saw. The distraction is performed by a build-in motorised system in the rod activated by an external remote control. It has advantage of small wound and comfort. The distraction forces are more direct. Initial reports are good at present. However there are potential drawbacks and problems. First, it violates and permanently destroyed the intramedullary bone and circulation. Second, infection and mechanical failure of the device will be a major disaster. The present modified Ilizarov method produces good lengthening results as reported by Ilizarov 70 years ago. It has become more refined and hopefully more comfortable for patient. Multiple wire and pin tract scars are still a drawback. The gain in stature in this group of patients had obliviate this cosmetic effect into a small blemish.

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