

EXTENSIVE LENGTHENING IN ACHONDROPLASIA

22 YEARS' EXPERIENCE

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(1) Common Dwarf, (2) and (3) Achondroplastic Dwarf, (4) and (5) Achondroplastic Dwarf, (6) Normal Native

From a photograph taken more than 100 years ago in India. Different types of dwarfism shown in comparison to a normal adult. The two on the right are Achondroplastic dwarfs.

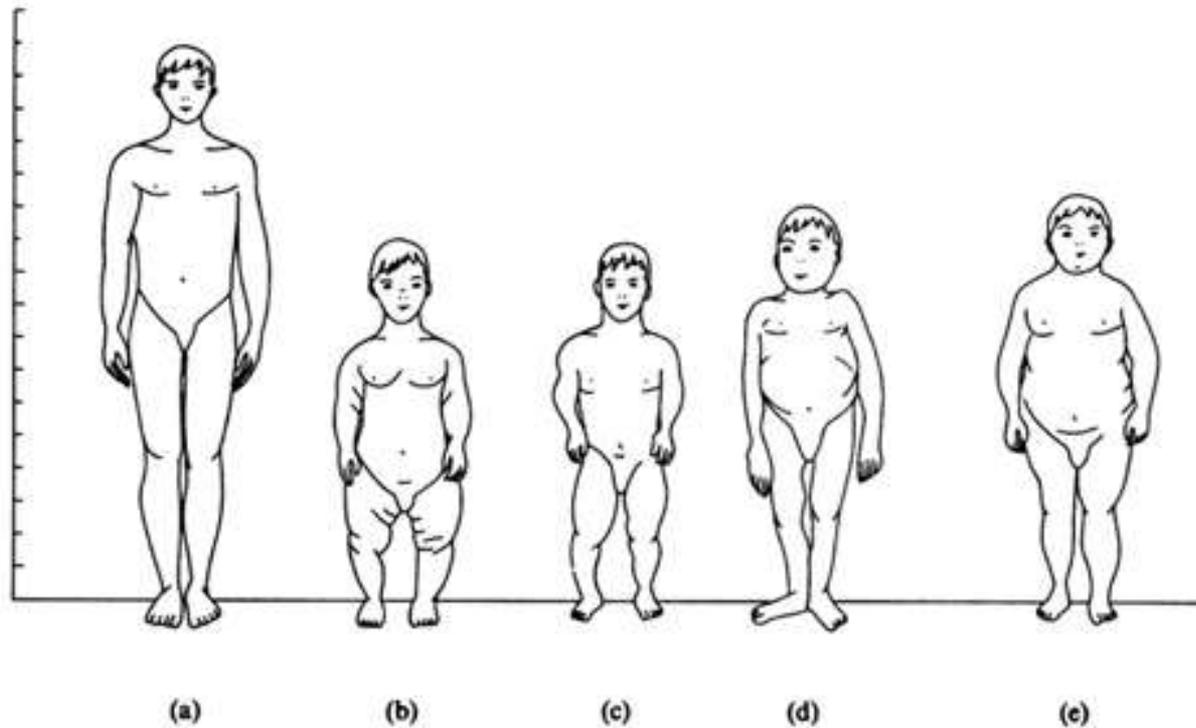


Figure 1. (a) Normal-sized adult. (b) Achondroplasia. (c) Pseudo-achondroplasia. (d) Spondylo-epiphyseal dysplasia congenita. (e) Hypothyroidism.

Achondroplasia is the commonest form of dwarfism with features that are hard to miss.

ACHONDROPLASIA

- ✘ Commonest form of short limbed dwarfism
- ✘ Incidence: 1 in 25,000 births
- ✘ Trunk normal — not detected on sitting
- ✘ Upper Segment : Lower Segment ratio **1: 0.6**

FACIAL FEATURES

- ✘ Frontal Bossing
- ✘ Depressed bridge of Nose
- ✘ Poor midface
- ✘ Large head



UPPER LIMB FEATURES

- ✘ Incomplete Extension of Elbows
- ✘ Trifid or STARFISH hands
- ✘ Short stubby fingers



LOWER LIMB FEATURES

- ✘ Bowlegs---Upper Tibial Varus
- ✘ Reduced rotation of Hips
- ✘ Cannot sit x-legged

LOWER LIMB FEATURES

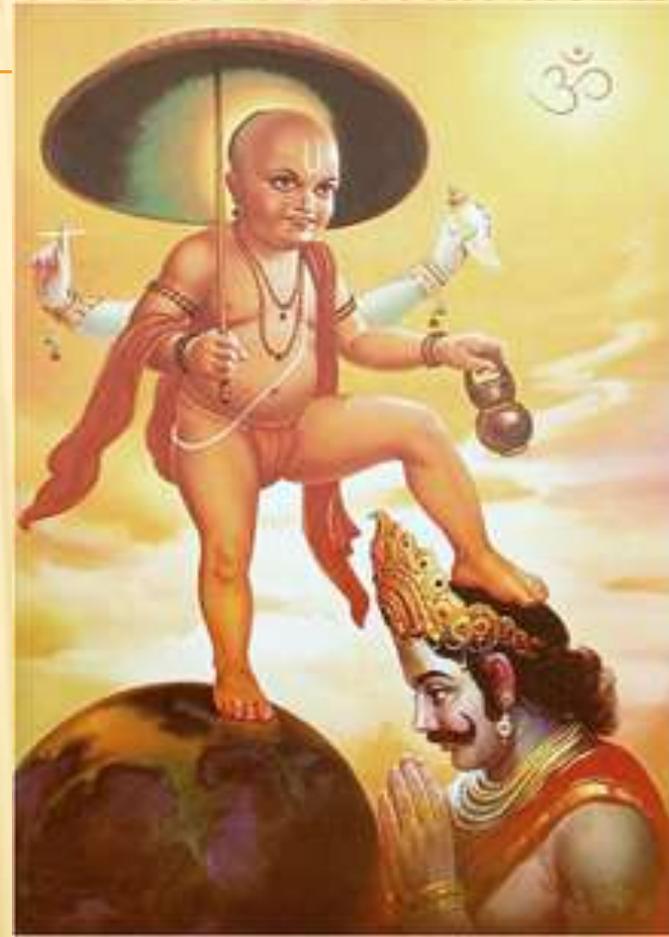
- ✘ Chevron shaped metaphyses
- ✘ Overlong Fibulae
- ✘ Upper Tibial varus
- ✘ Coxa Breva + high trochanters
- ✘ Lower Tibial Varus in older children



SPINE FEATURES

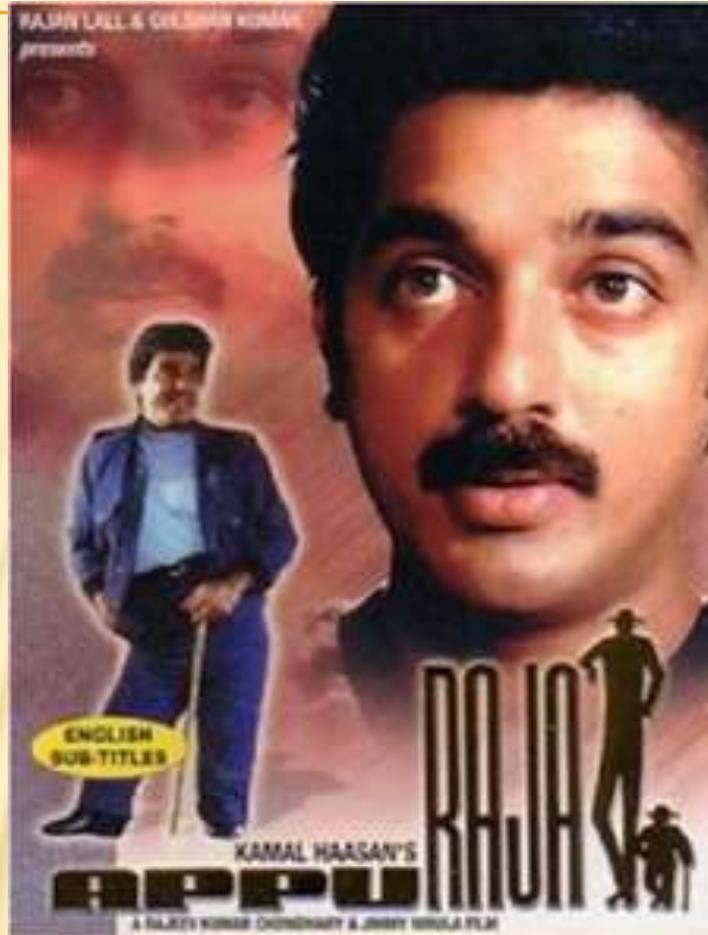
- ✘ Foramen Magnum Stenosis
- ✘ Cervical & Lumbar Canal Stenosis
- ✘ Dorsal Kyphosis
- ✘ Lumbar Hyperlordosis

VAMANA AVATAR OF LORD VISHNU-DWARF



Bali the king of Asuras with his penances wins over the 3 worlds. Vishnu comes in the “avatar” form of Vamana who is a dwarf and pleases the King and only asks for 3 steps. With one step he conquers the lands, with the second the heavens and when he has no place to put the third, Bali (who recognizes the Lord) asks to to place his foot on his head—thereby pushing him down in “Patala”

POPULAR CULTURE



Famous KamalaHaasan starrer (late 1980;s) showed the dwarf in a good light and brought this condition in the limelight of Indian audiences..

1990

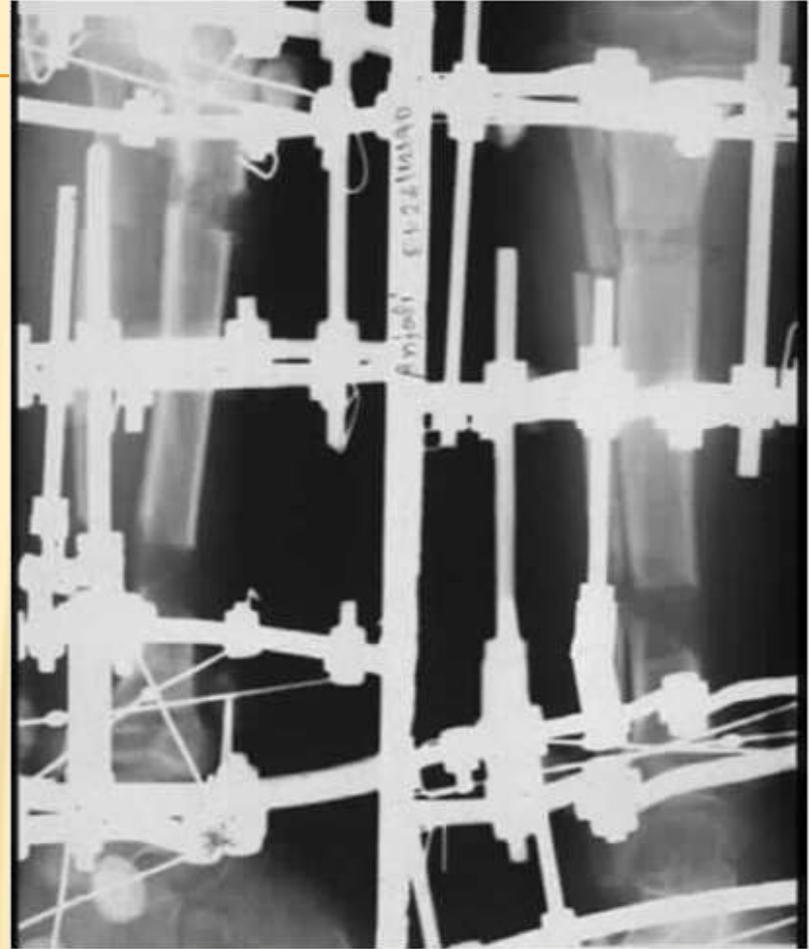


13 YRS. 3'11"



Our first patient from Bangalore, had severe bowing of legs and short stature.

DOUBLE LEVEL TIBIAL LENGTHENING



Double level tibial lengthening with a foot frame, allowed this 12 year old to walk. She developed a weakness of the dorsiflexion of hte Right foot due to paralysis of Lateral Popliteal Nerve. We stopped lengthening in Right Upper level to allow the nerve function to recover which it did in few weeks, We resumed lengthening after a repeat corticotomy.

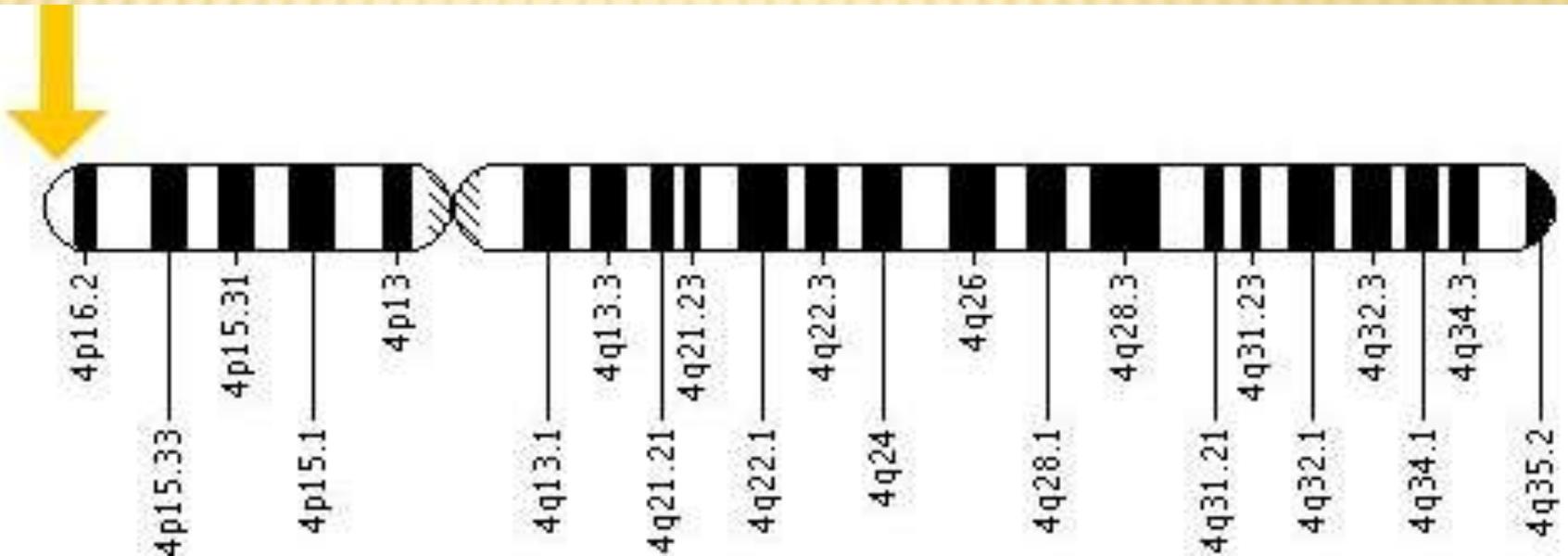
14 CM GAIN



She went on to get 14 cm length with no further problems. Her treatment duration was a total of 13 months and she was very happy with the result. She finished treatment about 21 years ago—the first in South Asia.

GENETIC BASIS

- ✗ Gly380Arg defect in the **FGFR3** gene
- ✗ FIBROBLASTIC GROWTH FACTOR RECEPTOR 3
- ✗ Cytogenetic Location 4p 16.3



BASIS FOR LENGTHENING

- ✘ Delayed maturation of Chondrocytes in the hypertrophic zone of Physis(growth plate)
- ✘ Reduced longitudinal growth of bone
- ✘ Length of muscles, nerves, vessels and other soft tissues normal—hence no resistance to lengthening.

Starting Early

- ✘ Age is on your side.....
- ✘ Children are small and easy to manage
- ✘ Possible to make them equal to their peers before joining nursery or Kindergarten
- ✘ Chances of improvement of facial features after limb lengthening.
- ✘ Lesser chances of developing lower tibial varus deformity.

5 YR OLD



SA came early and her parents were convinced that she had to go for lengthening.

LAT POPLITEAL PALSY & PREMATURE CONS



She had bilateral double level tibial lengthening as well . She too developed a temporary lateral popliteal nerve palsy and we slowed lengthening in the upper corticotomy area. The nerve recovered and we resumed lengthening with a repeat corticotomy.

11 CM GAIN
FN

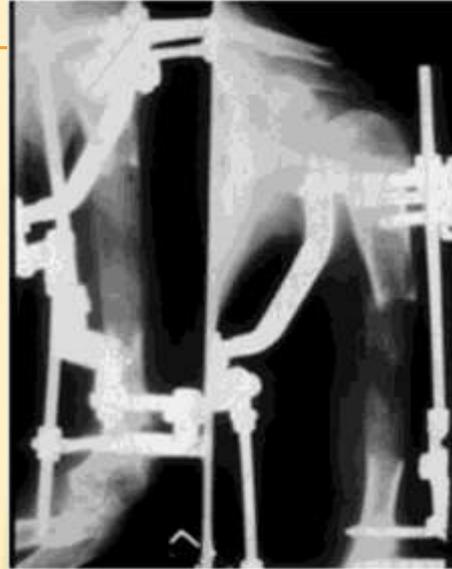
FULL RECOVERY OF NERVE

88% Lengthening



From there it was all smooth sailing and she went on to gain 11 cm height with full function retained in the joints.

HUMERUS LENGTHENING



Her Upper to Lower body proportions now have a big difference with the hands only reaching till the trochanters. This is not only cosmetic in appearance as it hampers the reach of the hands for hygiene and can contribute to dorsal kyphosis. She had a 9 cm lengthening of the humerus which restored her proportions.

CROSS LENGTHENING

TOTAL 33CM



She had a second stage Cross lengthening . The tibiae had repeat lengthening of only 4 cm but femur achieved 10 cm each. She had full movement in her knees. Her life has changed as she is now 4'10" –a very vivacious short normal!

× Bilateral Tibial Length

Foll by

× Bilat Femoral Lengthening



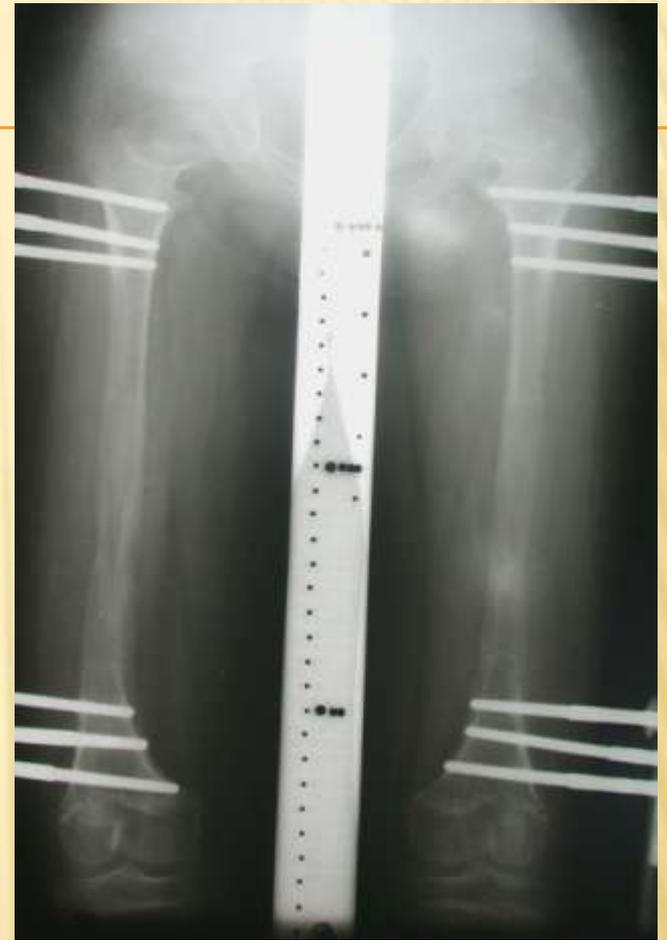
SH came to us as a 8 year old with severe bowing deformities and short stature.



We performed single level Tibial lengthening which gave her 10 cm length as well as corrected her bowing deformity. We also brought her Fibula down and tightened her Fibular collateral ligament which also helped reduce the varus.



At the end of the first stage her tibiae are straight she was walking very well



After two years she was ready for femur lengthening. We offered her lengthening with the LRS fixator which made it easy for her to walk during treatment. She achieved 10 cm length in the femora.

NOW A "SHORT NORMAL"



She is now a 16 year old who is considered a short normal. Leads a normal school life and is seeking admission in a medical school.



She has full function in her knees and almost full movements in her ankles which permit her to continue having classic Indian sitting patterns.

Starting Late

× More difficulties.....

12 YR OLD

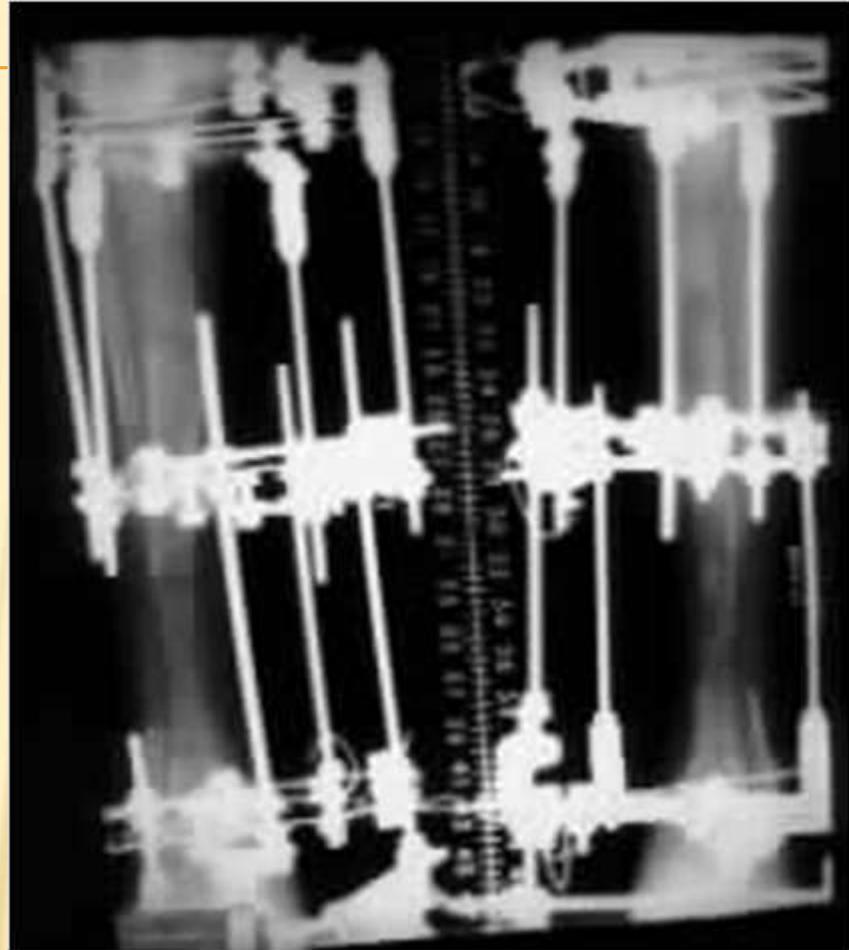
OBESITY & OBSTINACY



12 year old presented with severe genu varum, laxity and obesity. Genu Varum caused limited mobility which added to obesity.

15 CM GAIN

11 MONTHS



He had a Bilateral Double Level Tibial Lengthening which gave him 15 cm of increase in height. Treatment was harder due to his obesity and inability to walk much during treatment. However there were no serious complications at all. Duration was about 11 months.

15 CM GAIN



He did well and managed to achieve 6 inches. Had he started earlier, the parents could have thought about a repeat lengthening giving him more height.

HOW MUCH TO LENGTHEN

- ✘ Maximum safe possible...
- ✘ No pre-defined amount or Number to reach..
- ✘ Monitor the process..... Nerve & Joint Fn

% LENGTHENING

- × Tibia63% (15%---88%)
- × Femur.....49% (33%---65%)
- × Humerus.....50% (36%---60%)

AMOUNT OF LENGTHENING

- × Tibia 7 – 17 cm
- × Femur 8 – 17.5 cm
- × Humerus 8 – 10 cm

EXTERNAL FIXATION INDEX

- ✘ Tibia30.4 days/cm (21---54d)
- ✘ Femur.....25.4 days/cm (20--34d)
- ✘ Humerus.....28.5 days/cm (20--36d)

REGENERATE PROBLEMS

- ✘ Atrophic Regen. & BG.....0...*cf*.....
- ✘ Regenerate #.....0...*cf*..... 30% (Song)

PREMATURE CONSOLIDATION

- ✘ Incidence Tibial Lengthening8%
- ✘ Femoral lengthening.....10%

LIMB LENGTH DISCREPANCY

- × < 5 mm..... 3%
- × 5 –10 mm..... 4%
- × > 15 mm..... Nil

KNEE JOINT COMPLICATIONS

- ✘ Anterior Subluxation of Tibia1 knee
- ✘ Loss of ROM < 205%
- ✘ Loss of ROM > 200

AXIAL DEVIATIONS

✘ < 1015%

✘ 10 -156%

✘ > 153%

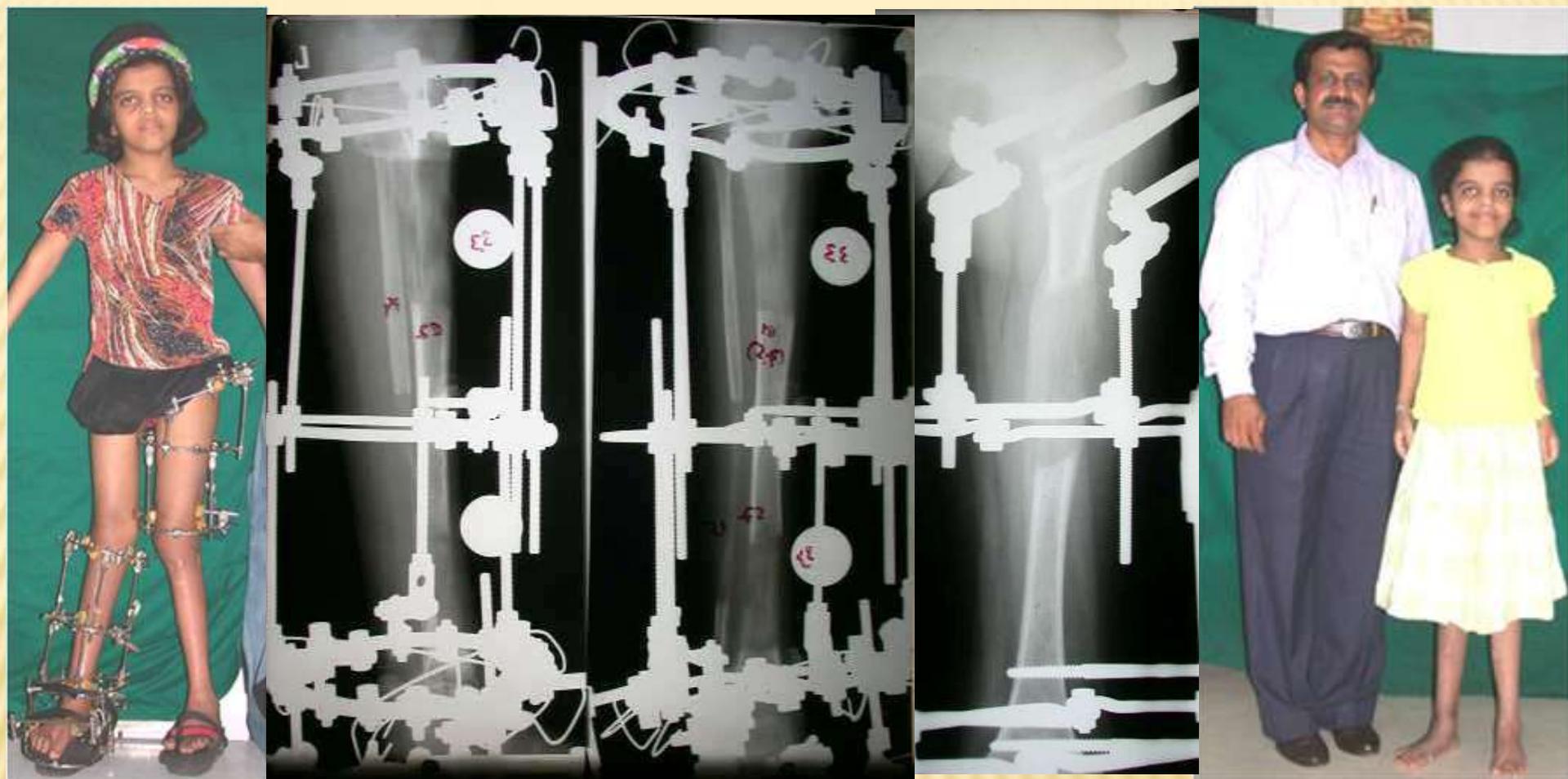
× Cross Lengthening

× &

× Humeral Lengthening



A doctor's daughter came at age 9 years with significant short stature. The father was sure that he wanted her to have the entire treatment in all 4 segments. Hence he chose to have Cross lengthening method. In the first stage the Right femur was lengthened at a single level giving 10 cm and Left Tibia at two levels giving 12.5 cm.



The second stage achieved similar results with her gaining significant height gain at 9 inches or 22.5 cm.



Humerus lengthening was added to restore body proportions. Humerus gained 9 cm in height.



Due to significant lengthening she went on to develop Valgus deformity in the Right tibia. This was corrected with an osteotomy using an LRS fixator. The Opp limb had a guided growth using 8 Plates to correct the valgus



Final result are straight limbs no deformities. More or less normal body proportions and she looks like a short normal.



Excellent Range of Motion is achieved with full function. She is now a Medical student.



15 yr old came with dwarfism from 1000 km away. The family was keen on the full treatment as well.



They chose a Cross Lengthening method. She achieved 12 cm in the Tibia with single level lengthening and more than 15 cm in the femur. Treatment lasted about 9 months



Second stage was started after few months and she went on to get a similar amount .



Her parents are pleased that she is now a short normal as she gained 27 cm in increase of height.

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