

Methods and Strategies in Limb Lengthening and Realignment for Skeletal Dysplasia

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INTRODUCTION

The recent availability of newer technologies of limb lengthening from Russia and Italy /1,2/ has led to a resurgence of interest in extended limb lengthening in dwarfing conditions /3-14/. The vast majority of limbs being lengthened are not skeletal dysplasia cases, but rather unilateral limb length discrepancies due to congenital anomalies such as fibular hemimelia. However, the lessons learned from these cases, and from our initial experience in stature lengthening for dwarfism do provide certain useful information. Indications for treatment for short stature are not well defined /15-20/. Moreover, not all short stature patients need or want lengthening. Concomitant angular deformities and joint abnormalities are usually more of a functional problem than the short stature. The lengthening devices provide useful means to correct angular, rotational and certain joint laxity problems /21/. Therefore, many skeletal dysplasia patients may benefit from the advantages of the lengthening technology used for non-lengthening indications. Numerous factors must be considered, including height, age, sex, etiology, trunk to limb disproportion, upper to lower segment disproportion, associated deformities, spinal pathology, functional limitations, psychologic factors, marital status, occupation, and socioeconomic status. Each of these factors may influence recommendations for treatment. In this paper, we describe our approach to the skeletal dysplasia patient who desires treatment for lengthening or malalignment, or both. Our experience is still relatively limited, due to our cautious patient selectivity. As our experience has grown, protocols have been changed to reflect the lessons learned. In the first half of this paper, we present our current concepts and methodologies. In the second section, we review our initial experience of lengthening in 20 skeletal dysplasia patients seen over a seven year period from 1987-1994. In contradistinction to other groups, we have found that these patients are not simply "lengthening" patients. Rather they benefit from undergoing simultaneous angular, rotational, and joint stabilization treatments. Furthermore, while the literature concentrates on achondroplasia and hypochondroplasia, we have found that also patients with pseudoachondroplasia and certain epiphyseal dysplasias may benefit from these techniques.

PREOPERATIVE CONSIDERATIONS

Age

With increasing age most individuals learn to accept the functional limitations imposed by short stature. Therefore, there are very few indications for lengthening for stature in adult patients over the age of 25-30 years. For maximum life year benefits, the lengthening should be performed prior to adulthood. Since the process of lengthening is so time consuming and temporarily disabling, it is best performed during childhood and adolescence, since this aspect of the life cycle has the fewest life commitments and dependencies. During adult life, major commitments, such as college, work, marriage, and children, make lengthening impractical.

Extensive limb lengthening requires prolonged bone healing times. Treatment time is related to bone consolidation rate. Bone healing is fastest in patients under 20 years of age. Younger patients have shorter treatment times and therefore large lengthenings are practical. Age is also a consideration in the amount of lengthening that is possible and advisable. Animal research has shown that extensive bone lengthenings in immature animals can inhibit natural growth by damaging the growth plate. Therefore, lengthening in children may be more safely accomplished in two or three small treatments rather than all in one large treatment.

Height

The primary goal of stature lengthening is increased height, but the surgeon's goals and the patient's goals may differ. Before lengthening begins, there must be a consensus regarding the goals of treatment. Function must not be sacrificed for height. Graphs and tables of normal height and of growth curves are available. Normal is defined as two standard deviations from the mean. This varies according to race and sex. The current indications for treatment are for patients who fall below the lower 2 SD, i.e., in the lowest 2.5th percentile. The goal of lengthening is to bring patients up to the lower limits of normal height. For Caucasian males this may be 160 cm and for females 150 cm. Having decided on a reasonable ultimate height goal, it is necessary to devise a strategy to achieve this goal. In the femur, the safe magnitude of lengthening ranges between 8-12 cm in one treatment. For the tibia and humerus the safe limits are 10-16 cm. These limits are mandated primarily by the soft tissues and the joints. Longer lengthenings require longer treatment times in the external fixture. Stature lengthening of 10-15 cm can be treated by tibial lengthening alone. This gives better cosmesis than lengthening of the femurs alone. Stature lengthening of more than 10-15 cm should be treated by lengthening both the tibiae and femora. After lower extremity lengthenings of greater than 15 cm, humeral lengthening should also be considered for functional and esthetic reasons.

If the proposed strategy is tibial lengthening alone, then tibial lengthening can be performed bilaterally simultaneously, allowing a four point crutch gait during

lengthening. When both femur and tibia are to be lengthened there are three possible strategies: 1) bilateral tibiae followed by bilateral femora; 2) ipsilateral femur and tibia on one side followed by the other side; or 3) contralateral simultaneous femur and tibia followed by the other contralateral pair.

Strategy 1 - [bilateral tibiae then bilateral femora]

This strategy maintains equal limb length throughout treatment. Since only one bone in each limb is being lengthened, double level lengthening of the tibia is possible. This substantially decreases treatment time. However, bilateral simultaneous femoral lengthening is difficult with ring fixators, due to inadequate space between the thighs. Monolateral fixators are more comfortable, but may not be strong enough to allow much weight bearing. Patients undergoing bilateral femoral lengthenings are best advised to use a wheelchair during most of the treatment, though they can stand with crutches for brief periods to transfer. This is our strategy of choice for most dwarfing conditions such as achondroplasia and hypochondroplasia (Fig. 1).

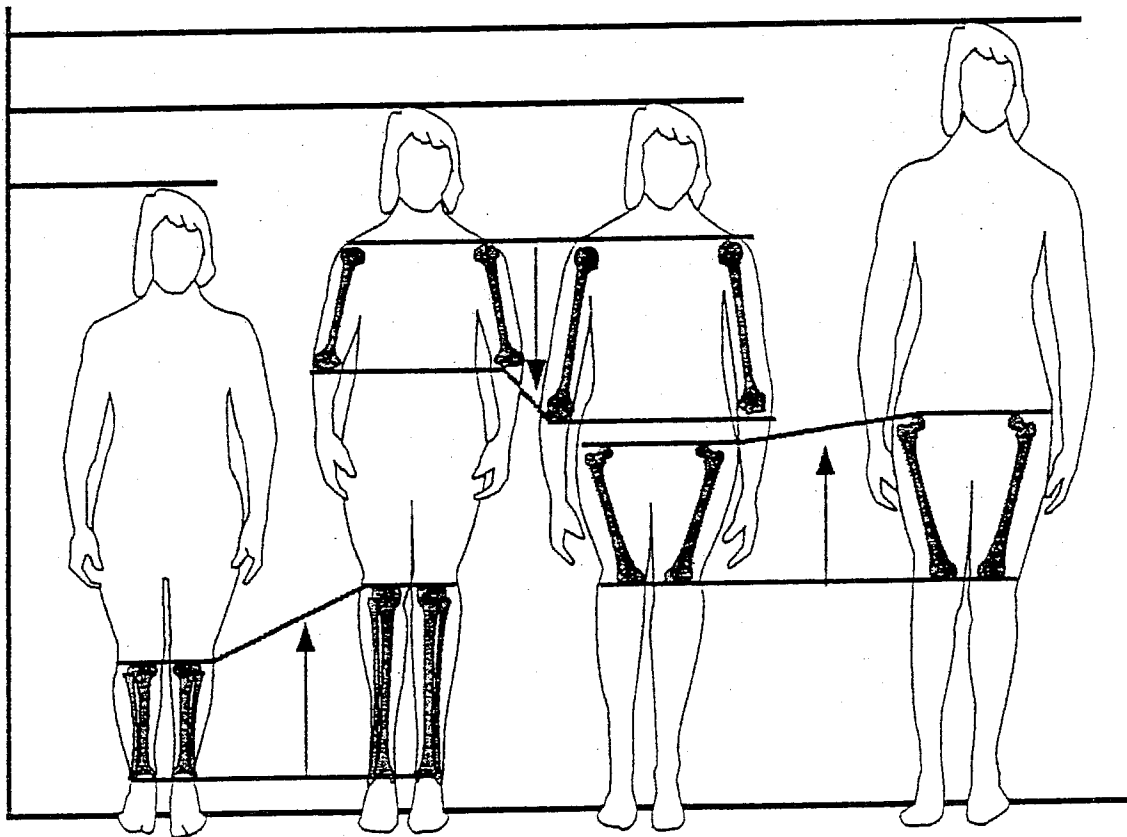


Fig. 1: Diagrammatic representation of strategy for staged lengthening in achondroplasia. First stage is 15 cm bilateral tibial lengthening at age 11. Second stage is 10 cm bilateral humeral lengthening at age 13. Third stage is 10 cm bilateral femoral lengthening at age 16.

Strategy 2 - [ipsilateral tibia and femur]

Since only one leg is operated on at a time, the patient can stand on the untreated limb comfortably and without difficulty. The biggest disadvantage is the creation of a leg length discrepancy. If for any reason (medical, social or financial) the patient has to stop treatment after the first leg, a leg length discrepancy will result. This is our strategy of choice when it is desirable to link the femur, tibia, and foot frames in order to protect the joints during lengthening, such as pseudoachondroplasia and multiple epiphyseal dysplasia (MED).

Strategy 3 - [contralateral tibia and femur]

This method is most useful if one is planning to use only the Ilizarov ring fixture. Bilateral simultaneous Ilizarov devices on the femur are very difficult for the patient, due to their bulkiness. There is, however, room for contralateral femur and tibia ring fixators. While the lengthening is bilateral a small interim leg length discrepancy is usually created because the magnitude of lengthening in the tibia and femur differ: more in the tibia than in the femur. We rarely use this strategy, as physiotherapy protocols may be confusing for the patient who is trying to simultaneously rehabilitate one femur and the opposite tibia.

Single vs double level lengthening

Double level lengthening can achieve a large amount of lengthening in a relatively short period of time by decreasing the healing time by nearly half. Due to the more rapid rate of stretch on the soft tissues, there may be more pain and a greater propensity for soft tissue related complications, such as contracture, joint subluxation, joint stiffness from increased joint pressure, and nerve stretch injury. Double level lengthening in the femur is usually not recommended. Tibial double level lengthening is more practical and tolerable. In the strategies described above, double level lengthening is often used on the tibial segment while single level lengthening is done on the femur, except for Strategy 2 where both femur and tibia are treated by single level lengthening to avoid three levels of lengthening in the same limb.

Humeral lengthening

Lengthening of the humerus is part of the strategy of stature lengthening when the stature lengthening is of large magnitude. Leg lengthenings less than 15 cm do not produce sufficient limb disproportion to require humeral lengthening. Leg lengthenings greater than 15 cm produce sufficient upper to lower limb disproportion that humeral lengthening is required for aesthetic reasons. When the lower limbs are significantly lengthened it becomes increasingly difficult for the patient to reach their feet with their hands. Furthermore, since part of the strategy of femoral

lengthening in achondroplasia is to reduce the lumbar hyperlordosis by either hamstring tightening or by proximal femoral extension osteotomy, there may be a resultant decrease in the hip flexion which would further impede the ability of short arms to reach the feet. The usual humeral lengthening amount is between 8 and 12 cm, done at a single level. In general, humeral lengthening is much more tolerable for patients than tibial or femoral lengthenings.

Timing of surgery

The question of when to start the lengthening process must be individualized. Factors to be weighed include the patient's age at first presentation, the psychology and motivation of the patient and family, the initial and expected height of the patient and the status of the patient's joints (related to their short stature etiology). Initial presentation at older ages limits the age strategy options.

Patients presenting before the age of 10

Achondroplasia and hypochondroplasia patients presenting before the age of 10 can be considered for lengthening between the age of 6-10. There are advantages and disadvantages to lengthening at this age. The main advantage is that a very large amount of lengthening may be ultimately gained (25-30 cm) by planning three shorter lengthenings rather than two larger lengthenings of the lower limbs. A 10 cm lengthening of both tibiae can be considered at a young age. Assuming adequate psychological maturity, it is less disruptive to lengthen a smaller amount at a young age than a large amount at an older age. A second advantage of operating on younger children is to correct and prevent bowlegged deformity in achondroplasia and valgus deformity in pseudoachondroplasia, epiphyseal dysplasia and chondroectodermal dysplasia. It is well accepted to correct these deformities at this age by osteotomy. The Ilizarov technique allows simultaneous accurate correction of angular deformities combined with lengthening. Ilizarov claims that anthropometric measurements performed before and after lengthening in young achondroplasts demonstrate a positive change in the facial and skull shape (less depression of the nasal bridge, more development of the base of the skull, etc.). He attributes these changes to endochondral ossification stimulation by humoral factors released during limb lengthening.

The disadvantages of stature lengthening in younger children are the small size of the bones and possible lack of patient motivation. The size of the bone makes double level lengthening impractical. However, it is probably unwise to do double level lengthening on young children because it puts excessive (harmful) pressure on the growth plates. The lack of motivation is probably the biggest obstacle to lengthening at a young age. Young patients with achondroplasia usually adapt quite well socially. The difference from their peers, while noticeable, is still not that large. They usually do not become self-conscious about their height until the adolescent years. One must be careful to avoid a situation where the motivation for

lengthening is coming from the parents who want their child to be "normal" and the doctor who wants to perform the surgery. Lengthening entails many potential complications and the treatment is especially stressful in a child who may not appreciate the rationale for lengthening. The child could become angry and depressed with serious resultant psychological ramifications. Therefore, lengthening (at any age) should be done only if the child is highly motivated. If the surgeon or parents have any doubt about the child's motivation, a psychological consultation should be obtained. Routine psychological consultation is practiced in many limb lengthening centers, in case unanticipated problems arise during treatment.

Patients presenting at age 11-19

By this age the difference between the child's stature and proportion and their peers is quite noticeable. It is now the child that makes the request for something to be done to normalize their height with their peers and to make them less noticeable so that people do not stare at them. The child wants to go through the growth spurt along with their peers. Other factors such as deformities also play a part in the motivation and rationale. Younger teenagers are usually in middle school or first years of high school. This is a convenient time to take off from school and is much easier than towards the end of high school or beginning of college.

Presentation age over 20

Young adults can still be considered for lengthening. The major limiting factors are the prolonged treatment time, inconvenience and financial considerations. Bone consolidation rates in patients age 20-29 are slower than those in patients under age 20. The longer the treatment time, the less the tolerance and the greater the problems experienced with the treatment. If the patient desires a large lengthening, it can only be achieved by repeated sessions of lengthening. This adds further to the intolerance and impracticality of the treatment. Before embarking on such a project, the young adult must consider the effect on college, work and family. Despite the best indications and desires, the surrounding life circumstances may not permit a patient to pursue lengthening treatment.

The strategy chosen may be scaled down according to the amount of practical treatment time. Usually the individual can afford to undergo the treatment only once. For such patients, lengthening of both tibiae makes the individual look taller since the knees appear higher. It also allows for more lengthening with fewer complications. Alternatively a strategy of two treatments may still be chosen while restricting the amount of lengthening to two short lengthenings instead of one large lengthening.

Hardware choices

Currently there are two major types of external fixture being used: circular or ring fixators and monolateral types. The monolateral fixators are less bulky, and

better tolerated. However, the ring fixators are mechanically superior and much more versatile for performing complex angular and other corrections. Regardless of the system used, the patient still has to wear the external fixture on the limb for many months until the newly formed bone is solid enough to go without support. In order to make this process more comfortable for the patient, several attempts have been made to make totally implantable telescopic bone fixation devices for lengthening. To date, none are available for use in the United States, though certain designs are currently being tested in Europe and Asia. Newer methods of lengthening with internal telescopic intramedullary rods will no doubt become available in the near future, making lengthening more practical in older age patients. In the meantime, we have devised a technique in Baltimore that uses combined intramedullary rodding with external fixation, to shorten the amount of time that the patient has to be in the external fixators by about half. Candidates for intramedullary lengthening rods must have little or no axial deformity. This method is reserved for the femur after age 14-16 and in the tibia after skeletal maturity.

ETIOLOGY

Achondroplasia and hypochondroplasia [Figs. 2,3]

These disorders are characterized by a disproportionality of limbs to trunk such that the trunk is almost normal in size while the limbs are short. The sitting height of these patients is almost normal while their standing height and arm span are significantly reduced from normal. Furthermore they manifest a rhizomelic disproportion so that the femur and humerus are proportionately shorter than the tibia and radius respectively. Both the height disproportion and the rhizomelic disproportion are variable to a lesser or greater degree with known standard deviations and growth charts. Hypochondroplasia patients do not manifest the spinal and facial deformities and are taller than achondroplastics. While 25-30 cm of lengthening may be required to bring a patient with achondroplasia to a normal height level, only about 20 cm is needed in hypochondroplasia. Consequently the humeral lengthening may not be necessary in hypochondroplasia since the disproportionality is less.

The most common limb deformity that needs to be addressed at the same time as the lengthening is genu varum with lateral joint laxity due to proximal overgrowth of the fibula. Ankle varus may also be a presenting deformity. Hip varus is often present, due to a varus neck shaft angle and overgrowth of the greater trochanter. Hip flexion deformity is also common. As part of the lengthening for stature program, most of these deformities should be simultaneously addressed.

Genu varum and ligamentous laxity about the knee [22,23]

The knee varus deformity occurs from relative overgrowth of the fibula compared to the tibia. This has been attributed to the size of their respective growth

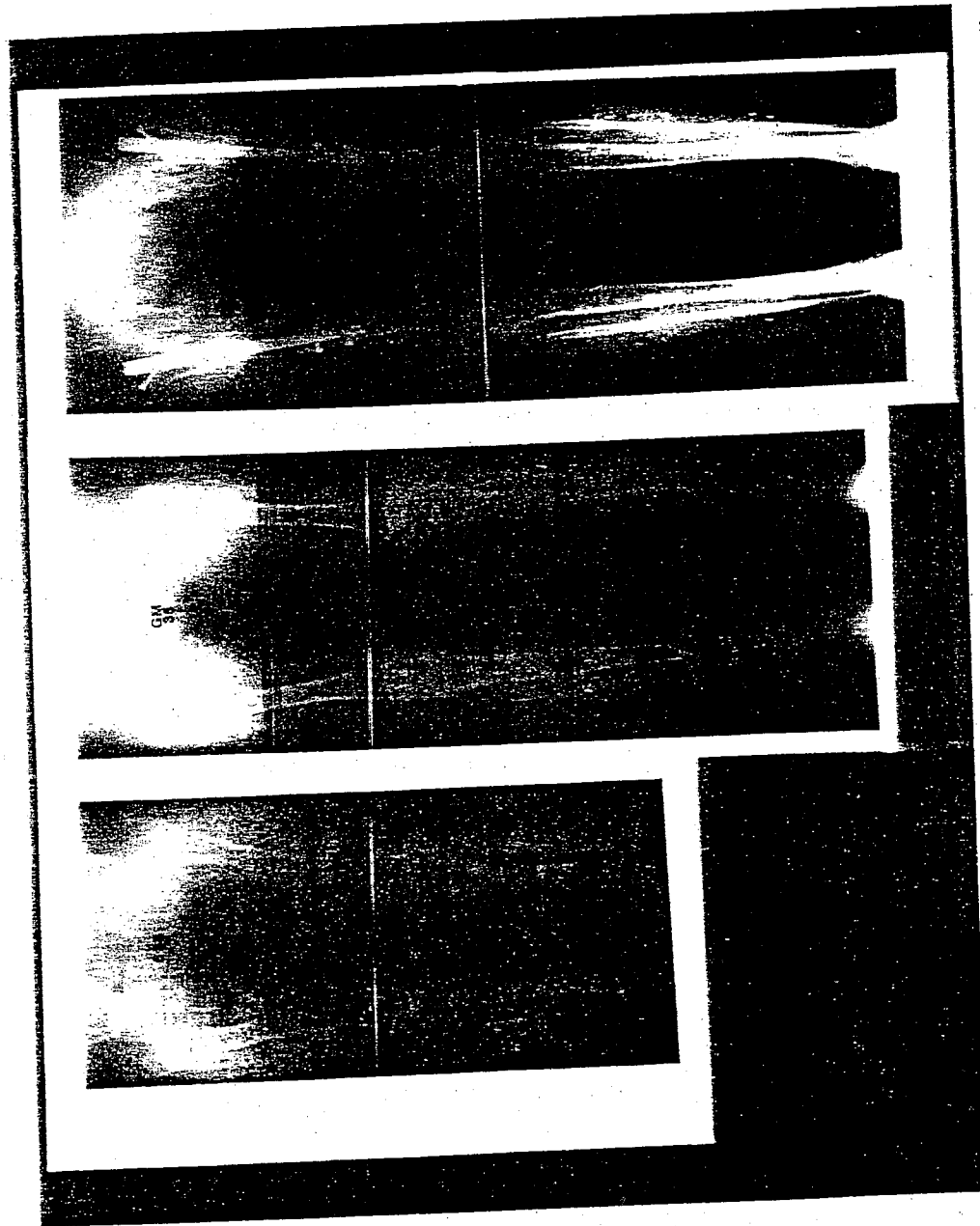


Fig. 2: Radiographs of staged lengthening in an achondroplastic dwarf. First panel: initial standing leg radiograph at age 12. Second panel: same view after double level tibial lengthening of 16 cm. Third panel: final view after 10 cm bilateral femoral lengthening over intramedullary nails. Total lengthening = 26 cm.



Fig. 3: Same patient as shown in Fig. 2. Left panel: standing at age 12 with father. Right panel: four years later, after completing 26 cm lower extremity lengthening and 10 cm humeral lengthening.

plates. Since the defect in achondroplasia is in endochondral ossification, its effect will be proportional to the cross-sectional area of the individual physis. The fibula has a small growth plate area compared to the proximal tibia, so it is less affected and therefore grows more normally than the tibia. Relative fibular overgrowth causes the leg to grow into varus. The insertion of the lateral collateral ligament moves proximally gradually creating more lateral laxity. Medial collateral laxity may also be present and should be checked using stress radiographs.

The varus deformity of the tibia can be corrected through the proximal lengthening osteotomy. The center of rotation of the deformity is usually juxta-articular and therefore requires an angulation-translation correction. In order to tighten the lateral collateral ligament during lengthening there are two approaches: 1) Double level tibial osteotomy with distraction of the proximal fibula. The fibula is fixed distally and in the midregion but is left unfixed proximally. The fibula is osteotomized only distally. As soon as the proximal tibial osteotomy is 2 cm distracted and the proximal fibula has descended 1-2 cm the distraction ceases at the proximal level but continues at the distal level. 2) Double level lengthening of the tibia and fibula are performed with fixation of the fibula at three levels. Near the end of distraction the proximal fibular wire or half pin is removed or backed out respectively and the proximal fibula allowed to descend. The latter option is the preferable one especially in children aged 12 and over. Medial collateral ligament laxity may not need to be addressed unless the patient is symptomatic. In such cases it is easiest to address this through double level lengthening to tighten the lateral and medial collateral together using an oblique osteotomy which begins proximal medial to the insertion of the medial collateral ligament and descends to exit distal lateral to the insertion of the patellar tendon. This osteotomy should only be distracted 2 cm in order not to overtighten the medial and lateral collateral ligaments. A second osteotomy for lengthening is used in the distal tibia and fibula.

An alternative approach to the fibular overgrowth and tibia vara problem has been resection of a large segment of the fibula (partial fibulectomy) to produce a fibular nonunion. Disadvantages are that it may lead to a valgus deformity, and it produces a nonunion in a normal bone. For most situations especially if lengthening is considered this is not recommended.

Distal tibial varus deformity

This deformity when present is centered at the juxtaarticular level and can be corrected through a supramalleolar osteotomy with angulation and translation. Since double level lengthening is usually the method of choice in adolescents, the distal osteotomy of the lengthening is used for the purposes of correcting the angle deformity as well.

Distal femur recurvatum deformity

This deformity is frequently present but does not need to be corrected unless it is of large magnitude (greater than 25° of knee recurvatum due to the bony defor-

stretch of nerves in the legs on spinal cord function. We have lengthened one such patient 12.5 cm in both tibiae without untoward neurologic effects.

Pseudoachondroplasia and epiphyseal dysplasias

Lengthening considerations

These conditions are characterized as having joint abnormalities, in addition to short stature. Early reports of lengthening in these conditions have included joint complications such as stiffness, joint deformation, and subluxation. For these reasons lengthening for stature has been considered contraindicated in these patients. The mainstay of treatment has been correction of angular deformities in order to prevent or delay late degenerative changes.

The severity of involvement in these conditions is variable. Pseudoachondroplasia (PSA) manifests the typical limb to trunk and rhizomelic disproportion as in achondroplasia but with many more deformities and with greater short stature. Multiple epiphyseal dysplasia (MED) shares many of the same joint deformities as pseudoachondroplasia but does not have rhizomelic disproportion. Spondyloepiphyseal dysplasia (SED) manifests the joint deformities but also presents with a short spine. Both SED and MED usually have less severe joint deformities than PSA.

The correction of limb and joint malalignment is the primary consideration in all of these conditions. Lengthening for stature can still be considered but to a more limited extent than for achondroplasia. Certain precautions must be maintained: the ankle should be decompressed for tibial lengthening by extending the fixation across the joint to the heel and applying mild distraction. The Achilles tendon should be lengthened percutaneously at the time of surgery. The knee and hip should be similarly protected for femoral lengthening by extending the apparatus across the hip and knee joints and by releasing the rectus femoris, fascia lata and adductor tendons. If humeral lengthening is performed protection of the elbow joint in the form of hinged distraction is generally not required nor are any soft tissue releases performed.

Using this approach we have achieved successful stature lengthenings of the lower limbs in such patients while preserving excellent joint function. In patients where the primary presenting problem is angular deformity, modest stature lengthening can also be carried out through the osteotomies used to correct the deformities.

Knee deformities

The typical knee deformities are juxtaarticular valgus and flexion of the distal femur and juxtaarticular varus and recurvatum of the proximal tibia. The center of rotation of these deformities is in the epiphysis of the two bones. Some component is at the joint line while some is in the physis. The other significant component of

the knee deformity is the ligamentous laxity and subluxation of the knee joint. These children have very lax joints but in particular there is marked laxity of the medial and to a lesser extent the lateral collateral ligaments. Ipsilateral osteotomies of the femur and tibia to realign the mechanical axis are the treatment of choice. The ligaments can be retightened by the methods previously described (oblique osteotomy of tibia starting proximal to the medial collateral ligament insertion and nonfixation of the fibula proximally). The oblique proximal tibia osteotomy is distracted 1-3 cm both to tighten the medial and lateral collateral ligaments and to achieve some lengthening for stature. The distal femur is osteotomized and its deformity is gradually corrected. Hinges are used to connect between femoral and tibial ring frames to protect the knee joint from increased muscle pressure during lengthening. In epiphyseal dysplasias, the unprotected joint will not tolerate the increased forces of distraction like a normal joint.

Hip deformity

The hip is usually positioned in flexion and varus. The trochanter may be overgrown. The femoral neck is short and the head may appear fragmented, notched or flattened (similar to Legg-Perthes disease). The natural history of these hips is to develop early degenerative changes and subluxation. If present, the acetabular deformity should be treated before femoral lengthening in order to prevent the hip subluxation from either natural growth or surgical lengthening and for coverage to increase the surface contact area. Secondly, the deformity of the femoral head can be corrected using a femoral osteotomy to correct varus. Flexion may be corrected partially or completely if the patient has more than 100° of flexion. If the patient has less than 100° the flexion should not be corrected since they won't be able to flex forward enough to tie their shoes, because they have short arms.

Ankle deformity

The ankle most commonly demonstrates varus and procurvatum but occasionally a valgus plafond is seen. The best correction of these deformities is by supramalleolar osteotomy. If the correction is performed acutely the tarsal tunnel may need to be released at the same time.

Mesomelic dysplasias

Mesomelic dwarfism usually presents with short tibiae and short forearms. The tibiae have varus deformities and lateral laxity. The forearms have a Madelung-like deformity of the distal radius. The severity of the short stature is mild in comparison to even hypochondroplasia. Treatment of the lower extremity varus deformity can be combined with bilateral lengthening of the tibiae. If fibular collateral ligament tightening is required it should be done as in achondroplasia near the end of the lengthening by removal of the proximal tibial pin. Upper extremity lengthening is not warranted.

Short trunk with normal lower limb length

This combination seen in Morquios, some spondyloepiphyseal dysplasias and following radiation to the spine (e.g. for tumors) requires special considerations. The disproportionality is caused by the shortened spine relative to the normal length legs. As with other dwarfing conditions, the standing height is affected but the more obvious finding is decreased sitting height. Lengthening the spine is not clinically done, although Ilizarov and others have successfully lengthened vertebrae in dogs and goats without creating paraplegia. Lengthening of the lower limbs increases both the stature and the disproportion. Modest leg lengthening of up to 10 cm does not worsen the appearance of the disproportionality very much but does address the stature problem. The patient should have a photographic representation of themselves made in order to decide whether the added height is worth the added disproportionality.

Short stature with normal proportions

Patients with certain chromosomal and endocrine abnormalities such as Turner's syndrome can fall into this category. Turner's syndrome patients are phenotypic females with XO abnormality and short stature. Growth hormone treatment has been successful in increasing height of Turner's patients prior to skeletal maturity. Once reaching adulthood, surgical lengthening becomes the only option to increase stature. The goal of lengthening for stature should be to achieve a height of 152 cm. This can usually be achieved with one lengthening of both tibiae. This produces a rhizomelic disproportionality as well as a trunk to lower limb disproportionality. These disproportions are minimally noticeable for lengthenings 10 cm or less. For goals of more than 10 cm, both femur and tibia should be lengthened. Bone consolidation may be delayed due to the postmenopausal state of these women despite replacement therapy. Calcium should be used in high doses during treatment (2000 mg/day). The tibial corticotomy should be kept as proximal as possible and as atraumatic as possible. Risk of late refracture (bending of regenerate) is also increased due to the underlying osteoporosis. Post removal casting or bracing should be used to prevent this complication.

Growth hormone deficiency patients should only be considered for stature lengthening if they are no longer candidates for hormonal therapy (e.g. past skeletal maturity). The other criteria are as for constitutional short stature.

Adrenal hyperplasia patients should also be treated with hormonal therapy. Their ultimate height depends on the development of precocious puberty. These patients are usually on steroid treatment. This must be considered preoperatively to ensure adequate steroid coverage. While a higher pin infection rate would be expected this has not been our experience with patients on steroids. The other criteria are as for patients with constitutional short stature.

Constitutional short stature

This is perhaps the most controversial of indications for stature lengthening. While not directly the subject of this symposium, it is nonetheless related. Many of these patients received growth hormone treatments during childhood, and may request surgical lengthening as adults. Many of the principles regarding evaluation and treatment are similar to those used for skeletal dysplasia patients. Since these patients are otherwise normal in every way, stature lengthening can only lead to functional worsening. Therefore the motivation for lengthening must be very strong to justify the risks. Motivational factors and psychology play a large role in the decision making for lengthening. These patients should undergo psychological evaluation to better understand their motivation and underlying psychological make-up. Many constitutionally short patients have psychosocial difficulties which they feel will be cured once they achieve greater stature. The short stature becomes the focus of the underlying psychologic disorders in these patients. In some cases, increasing the stature may help to resolve their problems. The decision to operate in these patients is difficult and controversial. Since functionally these patients are normal, they may be made functionally worse by limb lengthening. The decision should not be made in haste and all aspects of the process must be discussed and considered. There are three steps to the first evaluation: 1) history and physical; 2) patient education of theoretical and practical aspects of limb lengthening including strategies, methodologies, and commitments required; 3) patient education of risks, complications, results and costs.

The problem should be approached very clinically to start. The patient's height and proportionality should be measured. A full history and physical should be taken including evaluation of gait, running, jumping, squatting, muscle strength and range of motion of the limbs and spine. Even if the patient is obviously normal this complete examination should be carried out both for documentation and to stress to the patient that his/her complaint of short stature is being considered seriously.

Next, the motivation for stature lengthening should be explored in more detail. Signs of psychological problems, such as depression, anxiety, sexual dysfunction and sleep disorders, should be addressed. The socioeconomic status of the patient, including sexual life, marital status, family, occupation, and home situation, should be explored.

The patient should then be educated as to the technique of limb lengthening. This should include a description of the devices used, surgical technique, basic biology of bone and soft tissue regeneration. The discussion should include strategy, such as amount of lengthening possible, choice of limb segments, and order of limb segments. The expected duration of treatment and the follow-up and rehabilitation required should be frankly discussed.

At this point, the patient usually realizes that extensive limb lengthening is a very involved project. The patient can leave the consultation satisfied that they have evaluated the option and that it is not appropriate for them. However, if they are still interested, then the surgeon should provide a detailed description of all of the

risks and complications of the procedure. The possibility for worsening of function, as related to sporting activities, muscle strength and specific ranges of motion, should be emphasized. The risk of long term arthritis should be mentioned and the pain involved should also be emphasized and perhaps purposefully overemphasized. This discussion is enough to frighten off all but the most recalcitrant patients. However, if the patient is not dissuaded by the complications then the final discussion should be of costs. Since lengthening for stature in non-dysplastic individuals is considered cosmetic by most insurance companies, most will not cover the expenses of this procedure with occasional exceptions made for short stature due to iatrogenic treatments, such as mistimed epiphyseodesis, short stature leading to severe psychologic decompensation and short stature of such a degree that it interferes with function. If insurance will not cover the procedure, the patient must be made aware of the out-of-pocket expenses of treatment. Such a complete medical consultation as described frequently takes one hour in order to sufficiently educate a patient so that they can intelligently decide whether they are still interested in the surgery. After the patient's questions are answered, any literature available on lengthening and its complications should be given to the patient. The prospective patient should speak to previous patients who have experienced some complications so that the patient and the patient's family have a chance to see the issues from a patient's perspective.

For the small number of patients who successfully pass through these initial filters, a psychologic assessment is arranged. The purpose of this evaluation is to better understand any preexisting psychologic pathology and to better understand the patient's motivation for this surgery. Finally the psychologic assessment is used to better understand the family dynamics and support and the tolerance of the patient to stress and pain. The opinion of the psychologist is taken into account in the final decision making. Other team members, such as therapists and nurses, are also consulted.

Based on all these opinions and a strong willed and highly motivated patient, we occasionally consent to perform modest lengthenings for constitutional short stature. Even in a busy limb lengthening center, we find ourselves performing limb lengthening on non-dysplasia patients less than once per year. Most patients are screened out by initial telephone discussions and subsequent consultation. The screening process for skeletal dysplasia is similar to that just described, but requires a more extensive evaluation for concomitant medical manifestations of the dysplasia. Moreover, it is much easier to justify lengthening for dysplasia patients than for individuals with constitutional short stature.

BALTIMORE EXPERIENCE IN LIMB LENGTHENING FOR DWARFISM

Over the seven year period from 1987-1994 at the University of Maryland Center for Limb Lengthening, we have performed limb lengthening procedures on 20 skeletal dysplasia patients. Diagnoses included achondroplasia (11), hypochondroplasia (3), pseudoachondroplasia (3), multiple epiphyseal dysplasia (2), and

spondyloepiphyseal dysplasia (1). There were 13 females and seven males in this group. The goals of treatment in these patients were lengthening (20), angular deformity correction (14), and knee ligament tightening (5). It should be stressed that this preliminary update report must still be considered as a "work in progress". Most patients are undergoing a staged series of reconstructive procedures. To date all planned staged treatment has been completed on 9/20 patients, including 3/11 patients with achondroplasia, 2/3 patients with hypochondroplasia, 2/3 patients with pseudoachondroplasia, 1/2 patients with MED and 1/1 patient with SED. Mean stature increase has been 13 cm, with a range of 6-26 cm in the entire group.

Analysis of complications encountered may be divided into systemic, pin-related, bone, joint and nerve related. Systemic complications included one case of excessive blood loss requiring transfusion. This occurred in an achondroplastic dwarf teenager undergoing bilateral femoral lengthening over intramedullary nails. The blood loss from the bilateral IM nailing required post-operative transfusions. However, since then, we now ask all patients undergoing femoral nailing to donate autologous blood ahead of time. We have been able to avoid the need for homologous blood transfusion with this new protocol.

Superficial pin infections were ubiquitous in this series, as in all series of limb lengthening patients, averaging 1-3 pin infections per patient. In each case, oral antibiotics were sufficient to cure the infections. In one case, there was a ring sequestrum at follow-up. Bone complications included premature consolidation of corticotomy requiring re-osteotomy (1), deformity $>10^\circ$ due to bending of regenerate bone (3), osteomyelitis of the fibula requiring debridement (1), growth arrest (1), fracture after frame removal (3), and final limb length discrepancy >1.5 cm (1).

Joint related complications included severe knee contracture requiring extensive physiotherapy (3) and knee subluxation which resolved with brace treatment (1). Nerve related complications occurred in 12 nerves in 7 patients, including 6 achondroplasts. There were four bilateral and one unilateral peroneal stretch injuries that recovered with decompression, one unilateral peroneal direct drill bit injury that remains permanent, and two unilateral radial nerve stretch injuries that recovered (one was decompressed). The nerve stretch injuries generally appeared after 4-5 cm of lengthening. Our response to these injuries is immediate surgical decompression. After a brief pause in lengthening, we resume lengthening, even if initial recovery is incomplete. Using this protocol, final outcome has been excellent except in the one case of direct drill bit injury to the nerve, which remained permanent.

Overall functional results were satisfactory and all patients expressed that they were happy with the results of treatment. Most complications were treated satisfactorily, resulting in few lasting sequellae. Unplanned surgeries were required 23 times in order to treat problems and obstacles that arose during treatment. However, analysis of interim results shows a certain small but definable number of permanent sequellae: malunion of 10° (2), and permanent peroneal nerve palsy (1). In all three cases, patients are not unhappy with the result. The peroneal palsy patient walks without a brace and the two tibial malunions are satisfied with their appearance. In no patient was there a worsening of ability.

CONCLUSION

There is most certainly a "learning curve" effect, and we have learned certain lessons from our early patients that have served to make treatment of later patients go more smoothly. The ultimate question that the patient must answer is whether or not it was worth the effort, time and difficulties for the gains achieved. To date, we feel that our results in both lengthening and angular correction, and our results in correcting ligamentous laxity secondary to long term limb malalignment have been successful in offering selected patients with skeletal dysplasia satisfactory treatment options.

We continue to move forward cautiously in this area, bolstered by seeing the satisfaction of our skeletal dysplasia patients who have been lengthened, and feel grateful and satisfied with the outcome. Technological aspects of limb lengthening will surely improve in the next decade, making the process more comfortable and convenient for patients. Certain groups are already experimenting with totally implantable computer controlled battery driven telescopic intramedullary nails for femoral lengthening.

REFERENCES

1. Ilizarov G. Clinical application of the tension-stress effect for limb lengthening. *Clin Orthop Relat Res* 1990; 250: 8-26.
2. Paley D. Current techniques of limb lengthening. (Review) *J Pediatr Orthop* 1988; 8: 73-92.
3. Lavini F, Renzi-Brivio L, De Bastiani G. Psychologic, vascular, and physiologic aspects of lower limb lengthening in achondroplastics. (Review) *Clin Orthop* 1990; 250: 138-142.
4. Price CT. Limb lengthening for achondroplasia: early experience. *J Pediatr Orthop* 1989; 9: 512-515.
5. Saleh M, Burton M. Leg lengthening: patient selection and management in achondroplasia. *Orthop Clin N Am* 1991; 22: 589-599.
6. Molinari E, Mazzetti M, Peri G. The experience and the expectations of achondroplastic subjects during the period of surgical lengthening. *Basic Life Sci* 1988; 48: 445-446.
7. Weber G, Bregani P, Premoli F, Cassuffi MA, Turba F, De Angelis M, Chiumello G. Surgical lengthening of limbs in achondroplastic children: a medical and psychosocial program to select and treat patients. *Basic Life Sci* 1988; 48: 461-462.
8. Cattaneo R, Willa A, Catagni M, Tentori L. Strategies for limb lengthening in achondroplasia using the Ilizarov method - the experience of the hospital of Lecco, Italy. *Basic Life Sci* 1988; 48: 381-388.
9. De Bastiani G, Aldegheri R, Trivella G, Renzi-Brivio L. Lengthening of the lower limbs in achondroplastics. *Basic Life Sci* 1988; 48: 353-355.
10. Ascani E, Giglio GC, Crostelli M, Gasbarra E. Biomechanical problems, axial

- deviation and functional recovery in extensive limb lengthening. *Basic Life Sci* 1988; 48: 357-371.
11. Giglio GC, Pagnotta G, Caterini R. Neurological complications arising in bilateral lengthening of the tibiae in an achondroplastic subject. *Basic Life Sci* 1988; 48: 325-331.
 12. Mastragostino S, Bagliani FP, Boero S, Origo C. Techniques and results in extensive limb lengthening. *Basic Life Sci* 1988; 4: 333-351.
 13. Cattaneo R, Villa A, Catagni M, Tentori L. Limb lengthening in achondroplasia by Ilizarov's method. *Int Orthop* 1988; 12: 173-179.
 14. Aldegheri R, Trivella G, Renzi-Brivio L, Tessari G. Lengthening of the lower limbs in achondroplastic patients. A comparative study of four techniques. *J Bone Joint Surg* 1988; 70B: 69-73.
 15. Csapo M. Psychosocial adjustment of children with short stature (achondroplasia): Social competence, behavior problems, self-esteem, family function, body image, and reaction to frustrations. *Behav Disord* 1989; 16: 219-224.
 16. Shurka E, Laron Z. Adjustment and rehabilitation problems of children and adolescents with growth retardation: III. Bone diseases. *Acta Med Auxolog* 1976; 8: 215-221.
 17. Brust JS, Ford CV, Rimoin DL. Psychiatric aspects of dwarfism. *Am J Psychiatry* 1976; 133: 160-164.
 18. Moneymaker JM. The social significance of short stature: A study of the problems of dwarfs and midgets. *Loss, Grief and Care* 1989; 3: 183-189.
 19. Ablon J. Ambiguity and difference: Families with dwarf children. *Social Science and Medicine* 1990; 30: 879-887.
 20. Crandall R, ed. *Dwarfism. The Family and Professional Guide*. Irvine, CA: Short Stature Foundation and Information Center, Inc., 1994.
 21. Paley D, Herzenberg JE, Tetsworth K, McKie J, Bhave A. Deformity planning for frontal and sagittal plane corrective osteotomies. *Orthop Clin N Am* 1994; 25: 425-466.
 22. Paley D, Maar D, Herzenberg JE. New concepts in high tibial osteotomy for medial compartment osteoarthritis. *Orthop Clin N Am* 1994; 25: 483-498.
 23. Paley D, Bhatnagar J, Herzenberg JE, Bhave A. New procedures for tightening knee collateral ligaments in conjunction with knee realignment osteotomy. *Orthop Clin N Am* 1994; 25: 533-556.
 24. Lotti GM, de Pablos J, Gil-Albarova J, Cañadell J. Improvement of lumbosacral hyperlordosis after extensive femoral lengthening in achondroplasia. *J Pediatr Orthop Part B* 1992; 1: 55-58.
 25. Vilarrubias JM, Ginebreda I, Jimeno E. Lengthening of the lower limbs and correction of lumbar hyperlordosis in achondroplasia. *Clin Orthop* 1990; 250: 143-149.

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