

Limb Lengthening: Past, Present, and Future

David L. Rimoin, MD, PhD
Cedars-Sinai Medical Center
Los Angeles, California
Abstracted by
Robert M. Blizzard, MD

Introduction

In November 1990 at the National Cooperative Investigators Meeting of Genentech, Inc, Dr. David Rimoin gave an exciting presentation regarding the current and prospective status of leg lengthening to increase the height of individuals with short stature of skeletal etiology. This manuscript is an abstract of his presentation.

Limb lengthening has been a topic of increasingly developing interest over the past 2 years when reports first came out of Europe that it is possible to lengthen the limbs of chondroplastic children. Previously, the technique was used to correct limb asymmetry due to

polio, neurologic disease, and some congenital anomalies.

Wagner in Germany established one of the early techniques, which entailed breaking the bone surgically, performing an open osteotomy, cutting the periosteum, and using an external fixator and telescoping rod to stretch the tissues, which pulled the fracture site apart. Subsequently, he put in a metal plate, filled the area with chips, and it would heal. He operated upon achondroplastic individuals. He claimed that some needed up to 50 operations and that the complications were numerous. This technique has been abandoned for dwarfed individuals.

Ilizarov, an orthopedic surgeon working in a small institute in Siberia, was also one of the first to apply limb-lengthening techniques to dwarfs. In contrast to Wagner, Ilizarov utilized

“bloodless surgery,” as all the incisions were small percutaneous cuts. Utilizing little scalpels and chisels, he broke the bone percutaneously, put on circular fixators, and then gradually stretched the extremities. This limb-lengthening procedure involved slow, controlled distraction of the callus during its formation. Interestingly, histologic examination revealed that the new bone was very organized, longitudinally oriented, and that the organic matrix was capable of mineralization, which began to occur in the first few days. The new bone that started in the medullary canal ossified rapidly and underwent corticalization after stretching was stopped. Perfectly normal appearing bone was present once the procedure was completed. Complications of extended limb lengthening were significant, however, and only a

rare patient did not have at least 1 complication; and most had numerous complications. The real question was pointed out by Dr. Rimoin, who asked, "Are these complications worth what one gains?" Muscle contractures, neurologic compromise because of the stretch of the nerves, vascular complications, joint stiffness, problems at the pin site, and a variety of psychologic problems may occur.

A variety of techniques have been developed subsequently by Villarubias in Barcelona and a number of Italian investigators who use external distraction with only 1 bar rather than the circular fixator developed by Ilizarov. Dr. Rimoin stated that he now is convinced that these techniques are worth trying—particularly the Spanish technique. Villarubias lengthens both tibias initially, and both femurs are subsequently lengthened. This is in contrast to the techniques used by the Soviets and Italians, in which a femur and a tibia may be lengthened on the same or opposite sides. Villarubias's technique also differs in that distraction is started within a few days after the initial surgery rather than waiting a longer period. His technique also differs in that he does not permit weight bearing during stretching, as opposed to the weight bearing required by the Soviet and Italian procedures. Rimoin's observation is that there is much less discomfort and little pain in patients who utilize the Spanish technique. Villarubias also does prophylactic tenotomies while he is doing the initial surgery. This consists of splitting the edges of the tendons, thus preventing the contractures that have occurred with the other techniques. By his technique, the limbs are stretched approximately 1.2 mm per day. Patients are kept in the hospital for only a brief period (3 to 5 days) and are then allowed to go to school in a wheelchair.

It is of great importance not to disturb the blood supply, which

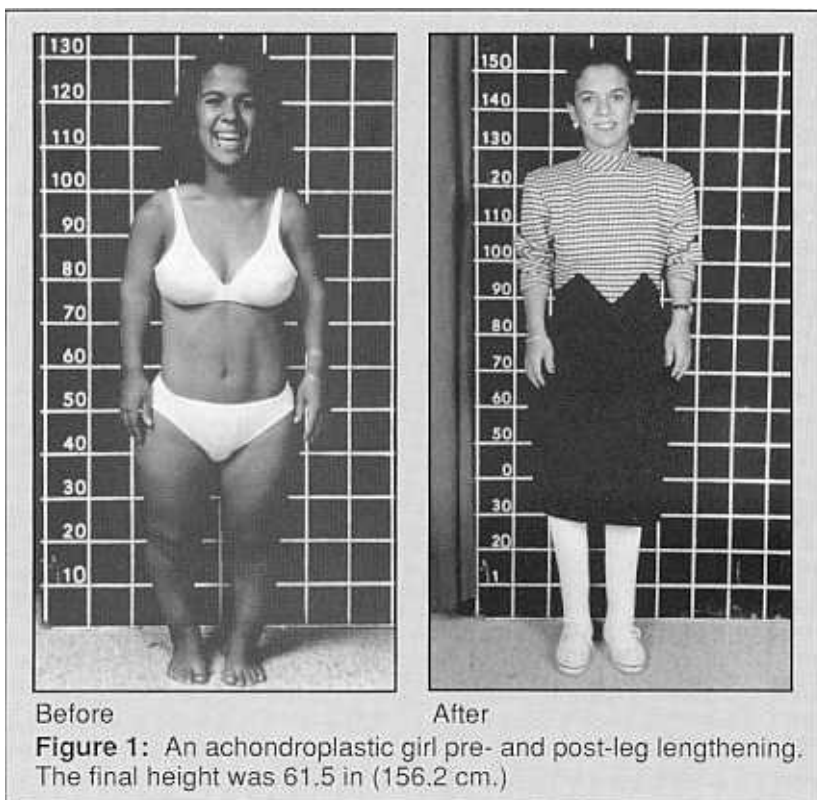
maintains the callus and allows it to heal rapidly. A few patients will have as much as a 30% reduction of ankle mobility and/or premature consolidation of the fibula, which may give obvious deviations of the extremity; about 2% have had to have repeat tenotomies. These complications are mild compared with those incurred with the other techniques.

Once the tibias have been lengthened, attention is focused on the femurs. Percutaneous tenotomy of the adductors is done early, and screws are put in the bones asymmetrically so that a rotational osteotomy is accomplished. A percutaneous tenotomy of the internal rectorus also is done, which relieves the tight muscles and tendons. No flexion of the knees is allowed during this time because the knee may be dislocated during femur stretching if it is placed in a fixed position. Patients are confined to wheelchairs.

Before initiating studies in Los Angeles, Dr. Rimoin visited Barcelona to observe the

technique of Villarubias. Subsequently, Rimoin sent 2 orthopedic surgeons, Drs. J. Isoaeson and W. Oppenheim, who are his collaborators, to learn the technique in Barcelona. As of the time of Rimoin's report in November 1991, 5 patients had been treated in Los Angeles. Two essentially had completed their leg lengthening. In Figure 1, the end result of a patient with achondroplasia who had such leg lengthening of both the tibias and femurs is demonstrated. This patient had a final height of 61.5 in. Utilizing this procedure, up to 12 in of increased growth can be anticipated. Dwarfs can be made into normal-sized individuals. Amazing to all observers is that the achondroplastic patients who have severe lordosis end up with essentially no lordosis when the Villarubias technique is applied. The coccyx is pulled down vertically, which reduces the lordosis markedly (Figure 2, page 6).

Dr. Rimoin states that the technique is still difficult to perform but it can achieve results. It must

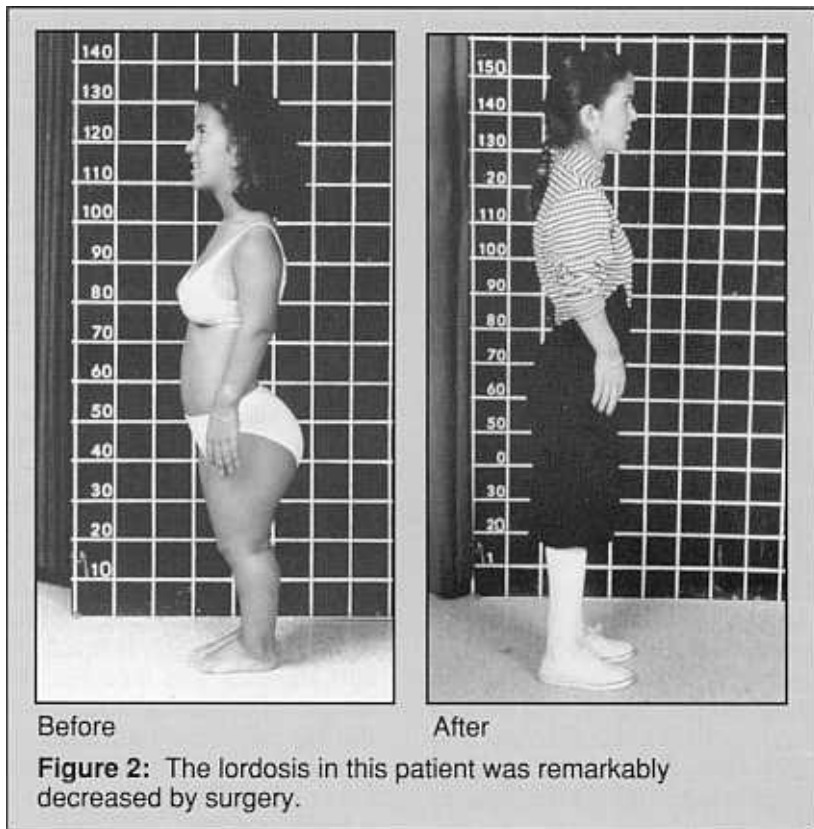


be done with a team whose participants are comfortable dealing with little people, that has broad surgical expertise, and comprises neurologists, geneticists, physical therapists, and psychiatrists or psychologists, all of whom are willing to be involved extensively.

Rimoin emphasized that one of the reasons that achondroplastic patients are such ideal candidates is that they have excess soft tissue in association with their short limbs. Because the soft tissues are in excess, and the blood vessels are long and tortuous, the soft tissues stretch readily and lengthening is primarily of the bone and not of the soft tissues.

What is the best time to perform the procedure? Ilizarov does it any time after 6 years of age. Villarubias does it any time after 10 years of age. Rimoin and his collaborators have decided upon 14 to 20 years. The emotional maturity that develops during the teenage period is important in helping the patients cope.

Rimoin thinks that in most instances it may be unwise to do the procedure before age 14 or 15. He emphasizes that it should be the child who makes the decision and not the parents, and



he emphasizes very strongly that the technique should be done in centers with extensive capabilities and experience by the team members. Dr. Rimoin pointed out that leg lengthening was less expensive in treating achondroplasia on an inch-for-inch basis than growth hormone

treatment in growth hormone-deficient patients. He emphasized that we all should be looking with great interest upon this technique, but that only a few should be trying it at this time.