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            Elio Ascani, and Victor A. McKusick

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HUMAN
ACHONDROPLASIA
A Multidisciplinary Approach

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PLENUM PRESS • NEW YORK AND LONDON
This volume is dedicated to Dr. Alexander Hollaender, General Editor of this Basic Life Sciences Series and Director of the Council for Research Planning in Biological Sciences, Inc., Washington, D.C., who sadly departed when our work was underway.

His advice, his experience, his unbiased criticism and his unrelenting encouragement would have been precious accessories to our task and we would have been proud to have had his final approval of the book. Unfortunately, I have lost a very dear friend and the entire scientific world now has a sorely felt gap.

Dr. Hollaender was a highly esteemed scientist of world-wide renown and an exceptional manager and organizer of scientific research, but above all, he was a man of honour with an acute and profound knowledge of mankind.

I am spokesman for many when I say: thank you, Dr. Hollaender, for having taught us how, where, and why to search for what sometimes seemed the impossible and for having left us a network of roads to follow in our continuing battle against suffering.

Benedetto Nicoletti
ACKNOWLEDGEMENTS

When it was decided to gather the material for this volume, its compilation was considered, erroneously, a straightforward procedure well within the routine of such matters. All those concerned with the preparation soon found themselves face to face with the tough reality of its being a complex and, often, difficult and frustrating enterprise; only sheer determination and a conscientious thought for the achondroplasts and their families who would benefit from the science contained within these pages spurred us on towards our target.

We feel it is our duty, therefore, to thank all those who made this book possible with their financial, practical and moral assistance:

- II University of Rome, Tor Vergata, Italy;
- The National Council for Research (CNR), Rome, Italy;
- The Ministry of Public Education, Rome, Italy;
- Merck Sharp & Dohme (Italia) S.p.A.;
- The Johns Hopkins University, Baltimore, USA;
- The staff of the International Centre for Skeletal Dysplasia, Towson, Maryland, USA;
- The staff of Plenum Press;
- Typists Elisabeth, Lidia and Lisa;

Last, but certainly not least, our deep gratitude goes to TRE EMME Congressi (Pisa) - without their patience and availability, without their indefatigable will-power and, above all, their magnanimous backing, very little could have been accomplished and this book would not have become a reality.
Benedetto Nicoletti with LPA delegate Ginny Brown underneath the "Hymn to Life", a wood carving by Bottega Michelangeli (Orvieto, Italy), which had place of honour in the auditorium during the First International Conference on Human Achondroplasia, which was held in November 1986 in Rome.
INTRODUCTORY REMARKS

We have come a long way in the last twenty-five years in our understanding of achondroplasia: its genetics, its differentiation from other simulating conditions, identification of the main clinical problems such as those arising out of cramping of the nervous system by the bones of the vertebral column and skull, and the prevention and treatment for some of these problems.

We have even further to go, however, in defining, for example, the precise nature of the biochemical defect so that tests for early prenatal diagnosis can be devised and attempts made at ameliorating the effects of that defect.

We have made great progress, I feel, in the area of social understanding of Little People, encouragement of acceptance in the workplace and mutual support in the achievement of a happy, satisfying and productive life. In the United States, this progress can be attributed in no small part to the Little People of America, Inc. Increasingly in other countries around the world, similar fraternal organizations of achondroplasts and other persons of short stature have served a marvelous function.

The psycho-social aspects of achondroplasia have come in for extensive discussions at this conference; these matters are fully as important as the genetics, biochemistry and surgery. The achondroplast must be made to feel his personal worth and encouraged to develop his full potential.

Victor A. McKusick
The First International Symposium on Human Achondroplasia held in Rome in November 1986 was a success. For the first time an international multidisciplinary team of physicians, educators, nurses, therapists, social workers, psychologists, short-statured persons, and patients and their parents convened for an in-depth discussion of medical and social concerns regarding achondroplasia.

It all started with the intention of Professor Nicoletti to convene a meeting of surgeons who perform Extensive Limb Lengthening (ELL) on achondroplastic individuals. The purpose of such a meeting was to compare surgical indications, techniques, results and complications with the hope that criteria would emerge for a prospective, controlled study.

It was clear from the outset that the approach of the medical profession to the problems surrounding achondroplasia was vastly different on the two sides of the Atlantic.

The shortness of stature of the achondroplast seemed to be the primary concern in Europe, and extensive limb lengthening was the prevailing treatment offered. Physicians in the United States were striving to find answers to the life-threatening and disabling impairments that affect achondroplasts during their lifetime.

Recognizing the valuable contributions of both camps, it was decided to expand the meeting to include communications by physicians and basic scientists with expertise in the multidisciplinary aspects of achondroplasia.

The situation became more complex when the social organizations of short-statured people of the U.S. (LPA), the U.K. (AARG), Australia and New Zealand, and Spain (CRECER), among others, wished to be represented. It was correctly argued that issues such as limb lengthening had deep social implications and short-statured persons should have a say.

The understandable apprehension of the organizers faced with participants of such diversity proved unfounded. The three-and-a-half-day meeting proved exciting for all. Most of the contributions were previously unpublished, many had never been delivered before. The days were full of surprises for even the most veteran in the field. The paramount issue of social acceptance was equally well presented: an eye opener for all.

Authors from the U.S., the U.K., Australia, France and West Germany
presented background information on the natural history and pathodynamics of the life-threatening and disabling complications. Cervico-medullary compression at the foramen magnum was presented as the major cause of death in infants. Symptomatic spinal stenosis at various levels was viewed as the leading source of disability in adults. Suboccipital craniectomy and decompressive laminectomies with or without spinal fusion were discussed. Attention was focused on the communication disabilities based on hearing loss.

Authors from Europe and the U.S.S.R. viewed their achondroplastic patients as non-participating, withdrawn, despondent, and without a chance in life. The patients' parents seemed to agree with them that "limb lengthening represented a chance for their children to feel more normal" and that it "increased their chances in life".

For most surgeons involved in ELL, this would be indicated for any short-statured person who wished to be taller, or whose parents wanted him to be taller. Older patients were generally discouraged from undergoing ELL, and children below five were not considered good candidates.

The techniques of limb lengthening were quite different from one another. Some surgeons lengthened as much as the patient would bear, while others limited their goals. Some allowed the patients to perform their own lengthening, while other surgeons were controlling the process themselves. Some were more open about reporting complications, although disagreement existed as to what should be called a complication.

The slides of the patient at the side of his parent before and after ELL were very impressive. Yet the clinical examination of five patients during the meeting was a sobering experience. It revealed the presence of significant complications.

The patients had been treated by different surgeons. Three had tibial, and two had tibial and femoral ELL procedures. Seven limbs had a footdrop due to peroneal nerve palsy. Two limbs showed telltale signs of ischemic changes. Five knees had marked posterior subluxation. Four patients had marked medial/lateral instability of the knees. The two patients with the femoral lengthening had an increased anterior pelvic tilt, not quite compensated by the lumbosacral hyperlordosis. This resulted in a forward pitch of the trunk. A history of chronic pin tract infection was present in two. The lengthened femur had fractured in one.

It is perhaps telling that the parents of these children did not mention any of these findings as complications. They were simply grateful for the height achieved. They find ELL an opportunity for their child and hope that the height gained will reduce the stigma attached to the appearance of dwarfism.

A program of extensive surgical lengthening of the limbs at the expense of such a high complication rate can only be justified if the "illness is more severe than the cure". In other words, the magnitude of social rejection, of the pressure to conform, can only be understood in terms of the price people are willing to pay to get rid of it.

The social implications of dwarfism are nearly always more serious than the medical aspects. Social rejection, real or perceived, is an
imprisonment of sorts where the victim becomes his own jailer and the sentence is for life.

Social acceptance of persons of very different appearance is still the essential element for the effective function of these otherwise normal individuals in society. Physical access should be assured by proper laws that provide for environmental adaptation of buildings, walkways, curbs, public restrooms, phone booths, elevators, etc. This should go hand in hand with access to higher education and employment opportunities. The "mass media" have been the "great educator" in the U.S. by portraying short-statured individuals in responsible social roles.

While these changes take place with time, ELL is here to stay in societies where dwarfism is objectionable. Yet it is not unreasonable for patients undergoing the procedures to expect respect for the integrity of their peripheral nerves and vessels, for the preserved integrity of their joints. They should equally expect freedom from the excruciating pain that characterized the experience for some. We believe the first ethical and moral responsibility of the surgeon is to his client, the child, who may not grasp the issues at the time.

It is also imperative that surgeons performing ELL familiarize themselves with the different syndromes enough to make the differential diagnosis between them and that they be aware of the pathologies that occur in the natural course of each.

Considering the prevalence and seriousness of the complications of ELL, the need for a controlled, prospective study is more imperative than ever. This is necessary, not only to establish proper parameters in order to prevent such complications, but to discern the effect of lengthening on residual growth, on the overall limb function, on symptomatic spinal stenosis, etc.

Generations to come may find our efforts to make dwarf patients taller by surgical distraction as barbarous and distasteful as we find the methods of the Great Inquisition. We can do better. We can solve our problems by knowing more about them, by a concentrated long-term effort at basic scientific research. It is the most costly, yet the only viable and lasting solution.

We want to express our gratitude to the Tre Emme Company for the organization of the First International Symposium on Human Achondroplasia and especially to Mrs. Shona Dryburgh for her extensive commitment to the publication of this work. We are proud to present this compendium of papers, intended as the single most important updated reference on the subject of achondroplasia.

Steven E. Kopits

The papers printed in this volume have been submitted by participants in the First International Conference on Human Achondroplasia; our intent is to divulge all that is known about achondroplasia and its complications. Moreover, for the first time there has been an attempt at investigating the various surgical remedies available for each of the
different complications, and much space has been devoted to the many
techniques and the results achieved, including an account of the lively
discussion which followed the session on extensive limb lengthening and
its problems.

The subject of extensive limb lengthening is of very recent origin and is
consequently the cause of much controversy; the aim of the conference was
to gather as much information as possible and to discuss the results for
an analysis of the benefits/risks involved.

From a personal point of view, even though I agree that mechanical
lengthening is not an answer to the problem of achondroplasia, being a
geneticist I cannot underestimate the importance of euphenics. My
daughter Donata, who is an achondroplast, has undergone this type of
surgery, which has increased her tibiae by 16 cm and her femora by 11 cm;
the 'price' entailed in reaching this height has been considerable, from
all points of view, but now she is much better adapted and is certainly
happier.

Taking into account the many other cases like hers, I think that,
ce once the methods have been standardised and the risks have decreased, it
would be opportune for orthopaedic surgeons in other countries to do their
utmost to make this type of surgery (which is to be considered an
improvement and not cosmetic) available to their patients. That is, until
science finds a way to cancel the effect of mutation.

Achondroplasia is, in fact, a complex syndrome which is essentially
disharmonic, stunted growth of the limbs - a situation which not only
causes many complications but also brings about severe functional and
psychological repercussions in the affected subject.

If society is expected to do all it can to accept all those who are
'different' in its midst, then I believe that the medical world should
undertake to reduce and, if possible, eliminate these differences which
interfere with the quality of life.

This is the invitation Donata and I wish to extend to all those who
are interested in the problem.

I am sincerely grateful to my dear friend Dr. Steven E. Kopits and to
Ms. Shona C. Dryburgh, who devoted themselves to the enormous amount of
work this required and without whom the volume would never have seen the
light.

Benedetto Nicoletti

The First International Conference on Human Achondroplasia had its in-
ception in the cultural and human encounter between Benedetto Nicoletti and
myself touching on the complex and multifaceted subject of achondroplasia;
we found common scientific and clinical interests which induced us first
of all to work in close collaboration and subsequently to want to organize
this meeting where every aspect of this 'Achondroplasia Planet' would be
treated and discussed.
It was a difficult and unusual experience, that of gathering in one meeting both scholars dedicated to basic research and clinicians coming from different specialities, all united by interest in this syndrome.

The very important contributions of sociologists, psychologists and re-educators could not be left out since their work takes on so much importance in improving the life conditions of short-statured subjects.

In addition to these specialists, who were speaking a scientific language, we wanted to give space to the patients themselves, to the families and to the associations which unite short-statured subjects in different parts of the world and which actively participated in the Conference. Our work of organization and choice of speakers of great renown, experts in the different aspects which we wanted to focus upon, availed itself of the valued collaboration of a man of great experience, namely, Steven Kopits, who, from very far away, contributed in a significant way to the successful outcome of the Conference. Setting up the state of advancement of the basic research on achondroplasia was the first target which we proposed to ourselves. An entire day of work was dedicated to the biological problems to underline the absolute necessity of integrating the experiences of the scientist and of the clinician; the first, a formulator of hypotheses from observation of the infinitely small, of what occurs before birth and of what happens before conception, the other, an observer of phenomena linked to the illness condition, ready to verify the truth and the possibility of transforming every suggestion coming from research into therapy.

In my clinical experience as an orthopaedic surgeon, though dedicated to infantile orthopaedics and to vertebral pathologies, I very often discovered 'black holes' in my culture when faced with illness where the orthopaedic pathology is associated and compared, in its natural history, with pathological conditions pertaining to other specialities. This is the eternal controversy of the cultural barriers connected with specialization, which on the one hand permit better assessment of the problems, but on the other hand make the physician lose overall vision of the patient's condition.

It is because of these negative experiences that we wanted to deal with all the pathological aspects of achondroplasia to discuss the internal, endocrinological, otoiatric, anaesthesiological, neurological problems and to give a single vision of the pathological picture, since, especially the orthopaedic specialist, who is faced in the achondroplastic with therapeutic problems of different degrees of importance, must not neglect a general evaluation of the patient before elaborating the often difficult recovery programmes, which require repeated surgery.

From the scientific sessions, it clearly emerged that each therapeutic act cannot be for its own purpose, but must always be finalized to the better functional recovery of the patient, in whom certain surgical interventions reply to a state of absolute necessity. Others (on the other hand) take on a role of necessity relative to improvement of the health condition. It is in this category of treatment that lengthening of the limbs belongs.

We are perfectly aware that to increase the height of an achondroplastic dwarf by 30 cm is not the equivalent of curing his
illness nor of making the patient a normal subject! Nevertheless, an experience personally started in 1983 with the Ilizarov technique has convinced us even more of the great physical and moral help which we give the achondroplastic subject by carrying out a great increase in his height.

In Europe, some experience has been gained in the past performing limb lengthening by the Wagner technique, but only the contribution of Ilizarov's theories, and the very rapid diffusion which there has been in the whole world of this cure system based on revolutionary principles has placed extensive lengthening of the limbs on the table of the therapeutic problems of achondroplasia.

To lengthen or not to lengthen? Two philosophies based on different experiences clash: the North-American negativist one, which my friend Kopits has skillfully summarized in his editorial, and the European positivist one, which has induced me, after learning directly at Kurgan the Ilizarov technique, to carry out 84 lengthenings of the limbs in achondroplastic subjects to date.

It is without doubt that a method which allows such exceptional results as doubling the initial length of a bone segment cannot be lacking in complications, especially in the initial phase of the experience.

With the precise purpose of not neglecting the incidence and the nature of these complications I wanted to dedicate a round table to the problems which emerged.

Nevertheless, the balance of the cumulative experience gained by the different groups in Europe which deal with this method is certainly positive because the complications tend to diminish as the experiences mature and the techniques are refined. To bring a subject whose average height is 125-130 cm to a height of 150-160 cm represents, however, a great help which one may give to an achondroplast:

The proposed solution is certainly temporary because lengthening is not the ultimate therapy for achondroplasia; but chemotherapy is not the definitive therapy in the oncological field; yet it is practiced since it is an efficient weapon in the cure of neoplasias.

This volume, which gathers the unpublished experiences reported in the congress, represents, in my opinion, a complete updating on achondroplasia.

Elio Ascani
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