Spinal deformity in neurological and muscular disorders

James H. Hardy, EDITOR

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edited by

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With 243 illustrations, including 2 full-color plates

The C. V. Mosby Company

Saint Louis 1974

Printed in the United States of America

Distributed in Great Britain by Henry Kimpton, London

Library of Congress Cataloging in Publication Data

Hardy, James H 1930-
Spinal deformity in neurological and muscular disorders.

1. Spine—Abnormalities and deformities. 2. Neuromuscular diseases. I. Title. [DNLM: 1. Neuromuscular diseases. 2. Scoliosis. WE735 H269s 1974]
RD768.H36 617'.375 74-616
ISBN 0-8016-2049-X

VH/GW/VH 9 8 7 6 5 4 3 2

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To all those children this volume
may help

Foreword

Virtual elimination or control of certain diseases such as tuberculosis, poliomyelitis, and infection of bone and joints has made resources and facilities available for an attack on a variety of other conditions. Scoliosis is one such condition.

Not too many years ago practically all patients with scoliosis were treated in a similar manner because the importance of etiology and curve patterns was not fully recognized, and relatively little differentiation was made.

Gradually, as different types of scoliosis have been separated and identified, we have been better able to prognose and select our treatment methods. For example, neuromuscular scoliosis has been divided into two major groups, myopathic and neuropathic. The differing prognoses of diseases within these categories have been better established, thus providing some selectivity as to which patients should be treated. Congenital curvatures have also been sorted out, and their characteristics are more predictable.

For some unknown reason lateral curvature of the spine has been considered different from anteroposterior curves. With increasing interest and knowledge about kyphosis and lordosis, principles learned from scoliosis are beginning to be applied to their management.

Surprisingly, adult sequelae of scoliosis received relatively little attention until recent years. Documentation of late effects of curvature of the spine has been made, but further studies are needed in this area. The heritable nature of idiopathic scoliosis is now beginning to be documented, and acknowledgment of familial trends has led to earlier recognition of scoliosis. Hopefully, we will learn more about the developmental history of scoliosis as physicians see more "early" cases. This may lead to modified or different treatment methods for scoliosis in incipient and early stages.

In the past, exercise programs, sleeping devices, casts, and an assortment of braces were tried in an effort to "compensate" spinal curves. As these methods failed, fusion, with or without attempted correction, was the principal alternative. Obviously no single method met with uniform success because all types of curves were dealt with in similar fashion. The Milwaukee brace has been successful in significantly reducing the number of curves requiring surgery, but the brace is currently being used in situations for which it was not designed and for which it is not suited. More work is needed to determine proper indications as well as limitations of the brace.

Although great advances have been made in the technical management of scoliosis, such as improved cast techniques, internal fixation with various types of spinal instrumentation, cephalopelvic traction, and other measures that allow early ambulation, it is only recently that much attention has been given to the total child and the impact of treatment on him and his family. A child does not function in isolation nor is he capable of independent action or decision. His environment, in contrast to the adult's environment, consists of his parents, siblings, school, etc. This environment has a continuing influence on his psychic and social development and must be considered and dealt with in selecting treatment methods in scoliosis. Psychosocial maturation in the growing child should constantly be kept in mind. These considerations demand close coordination of treatment of his physical and developmental needs. Considerable emphasis is now placed on mobilizing the child in his normal environment so that he may come in contact with people and places without sacrificing good correction of his scoliosis.

This volume should be useful to all those dealing with the fascinating problems encountered in the study of scoliosis. To the physician who is not dealing with large numbers of these patients, this book will be useful in making more timely and appropriate referrals. To the expert in the field it brings together current thinking of other experts, and since the volume points out limitations as well as the extent of current knowledge, it should further stimulate interest in the many unsolved problems.

This book is sufficiently inclusive that it will provide an important benchmark of current thinking to which we can compare in the future, and this may ultimately become the greatest value of the volume.

The authors are to be complimented for their thoroughness, their objectivity, and their clarity of presentation.

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Preface

Physicians in contact with a large number of scoliosis patients have come to realize that many of them have neurological or muscular disorders. Critical examination of a child with "idiopathic" scoliosis, for example, frequently will reveal abnormalities requiring a change in diagnosis. Increased awareness, more thorough neurological evaluation, and the preservation of the lives of many children who previously would have died in infancy mean a greater number of these difficult problems. Advances in preoperative and postoperative care and surgical techniques now permit procedures that were previously unthinkable. These factors plus a widening interest in scoliosis have led to many publications relative to the spinal deformity not only from orthopedic surgeons but also from neurosurgeons, neurologists, pulmonary physiologists, embryologists, and geneticists. This book is an attempt to gather this scattered information.

This book is not intended to be a definitive text or a compendium of surgical techniques. Rather it is intended to give the interested individual a broad background and understanding of the multiple problems involved—from the initial neurological evaluation to the postoperative pulmonary problems of a paralyzed chest. The more common neurological and muscular disorders associated with spinal deformities are discussed in depth, with illustrative case studies. The less common entities are mentioned briefly, but a basic analysis and rational approach to their treatment is outlined.

It should be emphasized, although a detailed discussion of the technique of spinal fusion is not within the province of this book, that the problem of a spinal curvature in a multiply handicapped child is not solved once the fusion is solid. Hopefully it is just the beginning of his becoming a productive member of society. This is the goal to be attained; the techniques and methodology described herein are only steps in an attempt to realize that goal.

James H. Hardy

Spinal deformity in neurological and muscular disorders

Contents

chapter 1

Introduction and general aspects

JAMES H. HARDY

Neuromuscular scoliosis constitutes a small portion of the total number of spinal curvatures, but the incidence of scoliosis in neurological and muscular disorders is high. In the past the major cause of neuromuscular scoliosis was poliomyelitis, but now that this disease has been virtually eradicated in developed areas, less common afflictions of the spinal axis have become more prominent. Basic concepts learned from treating the paralytic spine of poliomyelitis can still be applied to treatment of other neuromuscular curves, but several factors separate this type of spinal deformity from idiopathic scoliosis. First, a specific cause can usually be found for neuromuscular scoliosis. Second, in idiopathic scoliosis the curve may progress, whereas in neuromuscular scoliosis both the curve and the disease may progress.

The nature of the factors working in a growing and developing child can present a challenge to even the most accomplished physician. With neuromuscular disease the spinal deformity usually appears earlier in life than the idiopathic curvature and progresses rapidly and may not cease progression at maturity. Progression of curves may end only when there is a mechanical block such as the ribs on the pelvis, or the patient becomes so debilitated that they are recumbent. All too frequently these children die from pulmonary complications directly related to their deformed chest and not from progression of the neuromuscular disease. In a study of 88 children with congenital spinal muscular atrophy, all deaths were from pulmonary causes and not from the neurological disease itself.[5]

Etiology

The cause of spinal curvature in neuromuscular disease is usually readily apparent. The pathological process can lie at any site in the spinal axis from the cerebral cortex to the muscle fibers themselves. Curve patterns fall generally into two categories. The first type is that associated with a diffuse symmetrical weakness or paralysis and a collapsing spine (Fig. 1-1). The curvature can involve the entire spine, pelvis, and lower extremities, and evaluation of such a curve must include

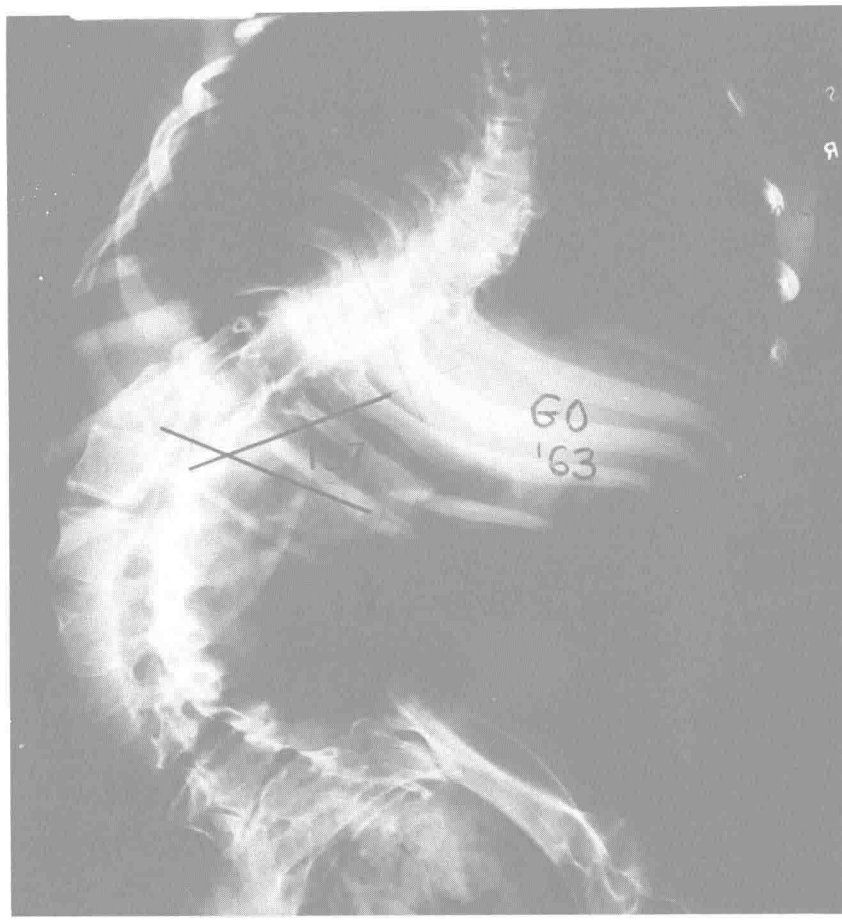

Fig. 1-1. Collapsing spine in an 18-year-old patient with congenital spinal muscular atrophy.

the entire body. The second type is related to a muscle imbalance. These curves can mimic idiopathic scoliosis so that the diagnosis of a neuromuscular disease is only made secondarily during a thorough evaluation of the patient with spinal curvature (Fig. 1-2).

Experimentally, dorsal root section in animals has produced scoliosis, with the degree of curvature directly proportional to the number of the dorsal root sectioned.[1,6] Some investigators thought there was a definite relationship between proprioception, balance, and the occurrence of scoliosis.[9] They created lesions in the brains of rats that subsequently developed scoliosis. However, other research attempting to reproduce these findings and to relate early scoliosis to proprioceptive defect has been inconclusive.[2] The application of these studies is not clear at present, but there is such a high incidence of spinal deformity in children with neurological and muscular disorders that the question arises whether all scoliosis does not have an underlying neuromuscular component. If the diagnosis of a neuromuscular disease is made, then an early scoliosis becomes a likelihood. The recognition of the curve is as important as the recognition of the disease, and often the curve may be more readily treated than the disease. Failure to appreciate a progressive spinal curva-

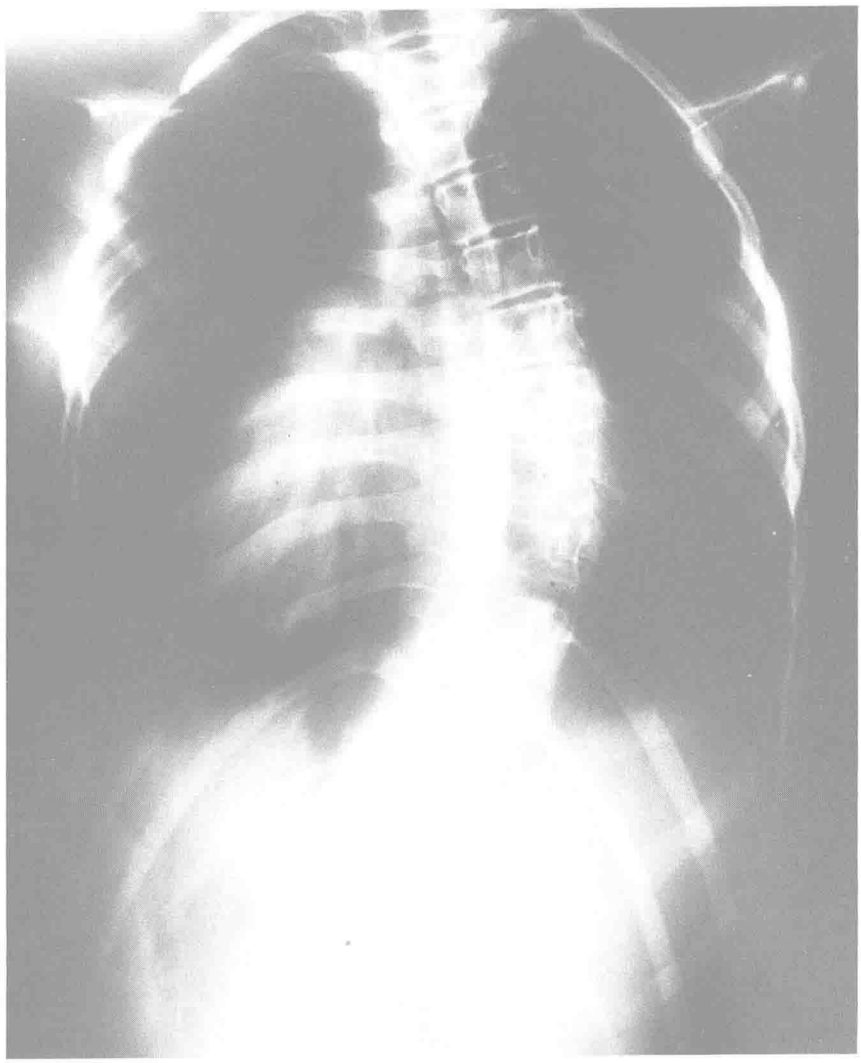

Fig. 1-2. The diagnosis of syringomyelia in this 13-year-old girl was made after scoliosis treatment had begun.

ture can lead to worsening of the child's general condition and even death from pulmonary compromise. It must be emphasized that curves appear early in life and often progress rapidly to a devastating degree.

Diagnosis

Diagnosis of the neuromuscular disease is important but not mandatory. Frequently, the prognosis can be deduced from the diagnosis, and therefore the diagnosis is more useful in deciding *which* individuals are treated rather than *what* is actually done. Occasionally a rapidly progressing spinal curvature will demand treatment before a final diagnosis of the disease has been made. There is much overlapping and confusion in the categorizing of the neuromuscular diseases, but several diagnostic steps are helpful.

Initially, a physical examination with a careful neurological study (as described in Chapter 2) can establish broad categories.

The electromyogram and nerve conduction studies are useful to separate afflictions of the central nervous system from those of the peripheral nerve or the muscle itself. It is difficult to get an adequate electromyogram in a young child, and the usefulness of this test depends on its intelligent interpretation.

Serum enzymes have taken an increasingly important diagnostic role in the past several years. The aldolase and creatine phosphokinase levels are both elevated in primary muscle diseases and are readily available laboratory tests.

A muscle biopsy can be most valuable. The method of taking the biopsy is almost as important as the interpretation of the biopsy (discussed in Chapter 4). A site should be selected in which the muscle is moderately involved and not one that is far advanced with only necrotic or fibrotic material. Electromyographic studies may be helpful at this time to locate a moderately involved muscle. A noncritical muscle should be chosen so that the patient's activity level is not compromised by the biopsy. Length and alignment of the muscle specimen must be maintained in the proper fixative so that artifacts are not introduced. A competent pathologist can be of great benefit in deciding what type of fixative to use and in selecting beneficial histological stains. Recently, histochemical and tissue enzyme studies, as well as electron microscopy have been used in the examination of specially prepared material. However, there have not been enough studies to standardize the method and obtain a base line.

The family history is especially significant in arriving at a diagnosis, since many neuromuscular diseases are hereditary in nature (discussed in Chapter 3).

The nature of the scoliosis may also be an aid in the diagnosis of the neuromuscular disease. For example, spinal curves usually appear late in Duchenne muscular dystrophy and ofter when the male patients are practically moribund. For this reason a spinal curve in a basically healthy, active young girl is a good indication that Duchenne muscular dystrophy is not a likely diagnosis.

Finally, it may be only after the clinical course of the disease has been observed for many years that the ultimate diagnosis will be apparent.

Treatment

Treatment for a child with neuromuscular disease and spinal curvature requires realistic and concrete goals. The effects of the disease should be separated from the effects of the scoliosis in an attempt to evaluate the improvement that will occur from the correction of the curvature. In its early stages the scoliosis may be positional or secondary to the disease process. However, with the passage of time the curvature acquires structural changes and becomes a progressive separate entity. Frequently, the correction or stabilization of the spine has an overall beneficial effect. For example, with the correction of trunk imbalance or severe decompensation, ambulation may be possible; breathing may be easier; and sitting may be more comfortable. In selecting a treatment program, it is valuable to separate general factors that are related to the child and the disease itself from specific factors that are related to the correction of the spinal deformity.

The general factors concern the overall ability of the child to withstand and understand treatment. The diagnosis of the disease and its prognosis now are im-

portant. The general condition of the child, such as whether he is bedridden or a walker, if he is cachetic or obese, or whether there is an associated osteomalacia, all would have an effect on the overall program and method of treatment. Frequently, other systems are involved in the disease process. Cardiomyopathy occurs in Duchenne muscular dystrophy. Of children with a congenital scoliosis, 18% have a congenital anomaly of the genitourinary tract.[7] In the skeleton, pathological fractures, Charcot's joints, and bone atrophy may occur in addition to contractures and dislocations. If nonoperative forms of treatment such as Milwaukee bracing are planned, the patient must be mature enough to understand and cooperate with the program. Protective sensation is also a consideration in deciding the type of cast, brace, or procedure that is going to be used. Nevertheless, the quality of life is important, and often this can be improved even though the quantity is unchanged.

Specific factors that influence the curvature involve structures beside the spine. The extraspinal factors include pelvic obliquity secondary to hip or other joint contractures, sitting balance or inability to balance, leg length discrepancy, and a dislocated or subluxing hip that is either congenital or paralytic. All these influence the foundation on which the spine rests.

With regard to the spine, the magnitude of the curvature and the location of the curve, especially if the pelvis is involved, are most important. Associated congenital malformations may also be present. Flexibility of the curve and the age of the patient will influence the treatment program. The main point is that the spinal curvature is only one facet of the overall problem. The approach to the spinal curvature should be coordinated with other considerations, including physical therapy, inhalation therapy, contracture release, extremity bracing, and other orthopedic and nonorthopedic procedures. Two big "Gs" are responsible for progression of such curves—*G*rowth and *G*ravity.

Nonoperative treatment

In a young infant, treatment of the scoliosis is difficult, but a "three-point pressure" type of brace can be used (Fig. 1-3). However, with a severe "floppy" child, this type of brace may not be possible, and a passive supportng device will be necessary (Fig. 1-4). To aid the child in obtaining the sitting posture and to prevent significant delay in this stage of the child's development, a more secure antigravity type of device is necessary. This can be a sturdy corset (Fig. 1-5) or a removable body jacket if the curve is mild and flexible and can be held under 20 degrees. If the scoliosis shows beginning loss of flexibility and a tendency to progression even when using the jacket or corset, a more sturdy body support or a Milwaukee brace can sometimes be used (Fig. 1-6). Ordinarily the use of the Milwaukee brace requires normal sensation, a degree of muscle coordination for exercising, and the intelligent cooperation of the patient. However, the Milwaukee brace has been able to manage curves in which two of these factors were present in a normal degree and the third was not absent completely. The most frequent cause of failure in the Milwaukee brace was the application of the brace when the curvature was too far advanced. If the curve is at 40 to 45 degrees, both bracing and spinal fusion enter into treatment considerations. When the deformity is over 60 degrees, spinal fusion is almost a certainty if the curve is progressing, but the brace may still be used as a temporary holding device (Table 1). In selected cases it may be possible to prolong