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Respiratory and Cardiovascular Functions in Scoliosis and the Principles of Anesthetic Management

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THE AIMS of this review are to provide a pertinent and succinct review of the etiology, and the respiratory and cardiovascular pathophysiology, of scoliosis, and to draw attention to the problems of anesthetic management of patients with scoliosis and the associated neuromuscular disease processes.

The abnormalities of respiratory and cardiovascular function in scoliosis include reduced lung volumes and compliance of the total respiratory system, arterial hypoxemia, which may be associated with hypercapnia, impaired chemical regulation of ventilation, and increased pulmonary vascular resistance. In addition, where scoliosis is associated with neuromuscular disease, these abnormalities are complicated by neural and muscular impairment of gas exchange and inadequate defense of the airways. Since the abnormalities of respiratory and cardiovascular function and their prognoses are dependent on both the cause of the scoliosis and the related orthopedic features, it is essential to consider these aspects as well.

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Incidence of Scoliosis

As a result of the prevention of poliomyelitis, recent surveys reflect largely the incidence of idiopathic scoliosis. The incidence in a survey of 11,000 Edinburgh school children in whom scoliosis was identified by clinical examination and confirmed roentgenographically was 3.9 per 1,000 girls and 0.3 per 1,000 boys. The overall incidence was 1.8 per 1,000.[1] A higher incidence of 4.0 per 1,000 was found in a roentgenographic survey in North America[2] because of the ability to identify more mild degrees of scoliosis roentgenographically.

Orthopedic Features

Structural scoliosis is a complicated deformity with both lateral curvature and rotation of the vertebrae, together with deformity of the rib cage (fig. 1).[3] In the area of the major curve, the vertebrae and spinous processes rotate toward the concavity of the curve, and the disks and vertebral bodies become wedge-shaped, and are narrower in the concavity. In addition, on the concave side of the curve, the pedicles and laminae are shorter and thinner, and the vertebral canal is narrower. As a result of rotation of the vertebrae, the ribs on the side of the convexity are pulled backward, producing a prominent posterior angle. On the concave side, the posterior angles of the ribs are flattened, and anteriorly, these ribs are prominent (fig. 1). These structural changes are most prominent in idiopathic scoliosis.

Kyphoscoliosis, which is a combination of kyphosis and scoliosis, is rare, and although it is usually a congenital abnormality, it is also seen in progressive

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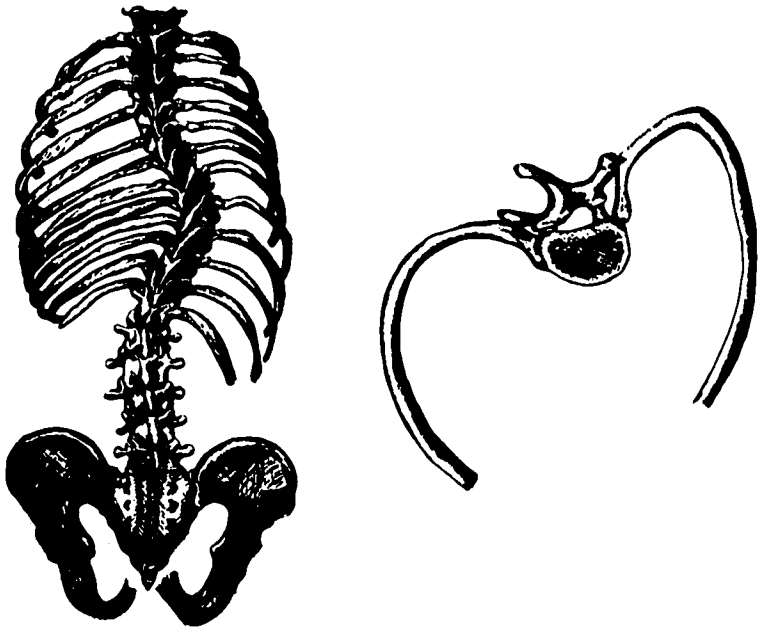

FIG. 1. Deformity of the vertebrae and rib cage in scoliosis illustrating the lateral curvature and rotation of the vertebrae, and the prominence of the posterior angles on the side of the convexity.

infantile scoliosis. The posterior rib hump in scoliosis may simulate kyphosis, but the term "kyphoscoliosis" should be restricted to disease processes that show kyphosis in addition to the lateral curve.

The severity of scoliosis is usually described by the angle of scoliosis.[4] Lines are drawn parallel to the upper border of the highest vertebral body and the lower border of the lowest vertebral body of the curve. To these lines, perpendicular, intersecting lines are drawn. The angle of curvature consists of the cephalad and caudad angles. The more severe the scoliosis, the larger are the angles. The deformity of the rib cage may be estimated from the size of the rib hump.

Diagnosis and Classification of Scoliosis

There are many etiologic groups in structural scoliosis (table 1). Diagnosis of scoliosis in an individual should include an identification of the etiologic factors and a survey of the family of the propositus. Several studies have reported a familial aggregation of idiopathic scoliosis.[5]

In order to establish the cause of the scoliosis, attention should be paid to examining the skin for *café-au-lait* markings and neurofibromas of von Recklinghausen's disease; pigmented areas of hair patches, which may indicate underlying congenital abnormalities; or ichthyosis, which is associated with degenerative neuropathy.

Clinical examination should include the respiratory, cardiovascular and neuromuscular systems, and should be supported by appropriate roentgenographic and other investigative data. Several studies have reported an association of congenital heart disease with scoliosis.[6–9] The neuromuscular examination should be relevant to the congenital, genetic, and degenerative neuromuscular diseases associated with scoliosis, and therefore should include examination of motor function, tendon reflexes, coordination, and segmental loss of sensory and motor function as in syringomyelia, diastematomyelia, or myelomeningocele.

Idiopathic Scoliosis

Idiopathic scoliosis is the most common type of scoliosis and, although the diagnosis is made by exclusion, it is characterized by many features. Idiopathic scoliosis has three well-marked periods of onset, which correspond to the peak periods of growth. It therefore is classified into three age groups: infantile (0–4 years); juvenile (4–9 years); and adolescent (10 years to skeletal maturity) (table 1). The thoracic and lumbar regions are the most frequent sites of the primary curve, and it is generally thought that the higher the primary curve, the worse is the prognosis.

Orthopedic Features

There are two types of infantile scoliosis: resolving and progressive.[10–12] In male infants, a significant proportion of the curves are progressive. These curves are usually thoracic, associated with a higher incidence of kyphosis, and, at maturity, are severe.[12] All of the 14 patients followed to maturity by Scott and Morgan[12] had severe deformities, the mean angle of scoliosis was 130°, and three patients died of respiratory and cardiac failure. There is a striking difference

between the incidences of progressive infantile scoliosis in Europe and North America. In an Edinburgh survey, the incidence was 1.3 per 1,000 infants 3 years of age or younger, whereas in North America, progressive infantile scoliosis is rare.[5] Although there is an increase in the proportion of affected first-, second-, and third-degree relatives, there are insufficient data for a clear genetic pattern to be defined.[5] Resolving infantile scoliosis does not usually increase beyond 30 degrees; it resolves spontaneously and does not require treatment.[4]

Adolescent idiopathic scoliosis is the most common; it is usually a right-sided thoracic curve and occurs more often in females. After onset, the curve progresses rapidly, involving seven to ten vertebrae. One curve in four will be greater than 100 degrees, and only one in three will be less than 70 degrees at the end of growth. Although it was once thought that there was little progression of the curve after cessation of growth, Collis and Ponseti observed that during a period of 25 years after completion of growth, adolescent idiopathic curves of 60–80 degrees increased by an average of 30 degrees.[13]

The familial incidence of adolescent idiopathic scoliosis is higher than that of progressive infantile scoliosis, and the data are consistent with a dominant inheritance with reduced penetrance.[5] Although most patients with idiopathic adolescent scoliosis are normal in other respects, in the Edinburgh survey 7 per cent also had mental retardation, congenital heart defects, and/or upper-limb anomalies. The same pattern of associated anomalies also occurs in infantile idiopathic scoliosis.

Juvenile scoliosis is a less well-defined group, and, although the Scoliosis Research Society recognizes juvenile scoliosis as a specific group, other authorities do not include this group.[1,4,14] The features of juvenile scoliosis and the problems in management have been recently reviewed by Tolo and Gillespie.[14]

Cardiovascular Function

Thoracic scoliosis from any cause may result in respiratory and cardiac failure. In the majority of patients described in case reports, death occurred before 45 years of age.[15] At autopsy, the incidence of right ventricular hypertrophy was high, and hypertensive pulmonary vascular changes were present.[16] The prognosis of cardiovascular function in idiopathic scoliosis, however, has not been rigorously determined, since in many of these studies either the cause was not established or the data for scoliosis resulting from several causes were pooled.[17,18]

A long-term study (50 years) on the cardiovascular and respiratory function in patients with untreated idiopathic scoliosis demonstrated that their mortality rate was twice that of the general population, and that the average age at death was 46.6 years.[19] Respiratory or right heart failure accounted for 60 per cent of the deaths. Of the living patients, whose mean age was 62 years, 47 per cent were incapacitated. However, in contrast, Collis and Ponseti's patients with untreated idiopathic scoliosis observed for 25 years showed no increase in mortality and morbidity.[13] The differences in morbidity and mortality between these studies may be due to the shorter period of follow-up in the Collis and Ponseti study.

It is postulated that several mechanisms contribute to the elevated pulmonary arterial pressure that is the result of increased pulmonary vascular resistance. First, in scoliosis, the number of vascular units per unit volume of lung is less than that of normal lungs.[20,21] This is caused by the impaired development, from birth to childhood, of the pulmonary vascular bed as a result of rib-cage deformity.[21,22] Second, in the regions of the lung compressed by the rib-cage deformity, the alveoli approach the volume of or become less than their residual volume. In these areas, therefore, blood flows in extra-alveolar vessels, which have increased resistance. Third, the inverse relationship between the pulmonary arterial pressure and the arterial oxygen

Table 1. Classification of Scoliosis

Idiopathic
Infantile, <4 years old
Juvenile, 4–9 years old
Adolescent, from 10 years old to skeletal maturity
Congenital
Spinal deformity
Abnormal spinal cord development
Congenital rib fusions
Neuromuscular
Neuropathic
Lower motor neuron, *e.g.*, poliomyelitis
Upper motor neuron, *e.g.*, cerebral palsy
Other, *e.g.*, syringomyelia
Myopathic
Progressive, *e.g.*, muscular dystrophy
Static, *e.g.*, amyotonia
Others, *e.g.*, Friedreich's ataxia
Associated neurofibromatosis (von Recklinghausen's disease)
Mesenchymal disorders
Congenital, *e.g.*, Marfan's syndrome
Acquired, *e.g.*, rheumatoid
Others, *e.g.*, juvenile apophysitis
Trauma
Vertebral, *e.g.*, fracture, radiation, surgery
Extravertebral, *e.g.*, burns, thoracoplasty
Secondary to irritative phenomenon, *e.g.*, spinal cord tumor

Table adapted from the Scoliosis Research Society[4]

tension or hemoglobin saturation, and the reduction in pulmonary arterial pressure with the administration of oxygen during respiratory failure,[23] are consistent with several arguments concerning the role of hypoxia in causing pulmonary hypertension. For simplicity, these arguments include: 1) a common cause for both the increased pulmonary vascular resistance and alveolar hypoxia, *e.g.*, the severity of the deformity; 2) local, regional, and general alveolar hypoxia due to ventilation/blood-flow maldistribution and hypoventilation, which causes reversible vasoconstriction and results in hypertensive pulmonary vascular changes.

Although the electrocardiogram provides a simple noninvasive tool in the preoperative evaluation of patients with scoliosis, electrocardiographic abnormalities indicative of pulmonary hypertension, as shown by right atrial dilatation (P wave > 2.5 mm) and/or right ventricular hypertrophy (R > S in V_1 and V_2), are late in appearance. Indeed, Shneerson *et al.*,[18] in a recent study of 40 patients with scoliosis from a variety of causes, observed hemodynamic evidence of pulmonary hypertension in ten patients, but noted that typical electrocardiographic changes of right atrial dilatation or right ventricular hypertrophy occurred infrequently. Shneerson *et al.*,[18] also reported that a prolonged interval between pulmonary valve closure and tricuspid valve opening (P_2-T_0) identified by phonocardiography was the most helpful noninvasive indicator of pulmonary hypertension. Recently, echocardiographic techniques have been used successfully to study right and left ventricular wall thickness and cavity dimensions in chronic pulmonary disease. This technique should provide a useful noninvasive method for assessing right ventricular structure indicative of pulmonary hypertension in scoliosis.

Respiratory Function

The most common pulmonary function abnormality is a restrictive pattern of the lung volumes. As illustrated in a recent study, the greatest reduction is in the vital capacity (VC) (mean per cent predicted 60.5 ± SEM 2.7). The mean per cent predicted total lung capacity (TLC) was 70.2 ± SEM 2.6, and the mean per cent predicted functional residual capacity (FRC) was 79.3 ± 3.2. The lung volumes and the compliance of the total respiratory system and of the chest wall are inversely related to the angle of curvature.[24] Several mechanisms may be postulated for the effects of scoliosis on the elastic properties of the respiratory system and on the lung volumes. These include the effects of scoliosis on the development of the thoracic cage, the direct effect of the vertebral and rib-cage deformity on the elastic properties of the respiratory system, and the effects of the deformity on the development of inspiratory and expiratory muscle forces. There are insufficient physiologic data to confirm which of these are the major problems.

Although Fishman and associates[17,23,25] concluded the major abnormality in gas exchange in scoliosis was general alveolar hypoventilation in which the lungs were normal but inadequately ventilated, the most common blood-gas abnormality is a reduced arterial oxygen tension (Pa_{O_2}) with a normal arterial carbon dioxide tension (Pa_{CO_2}).[17,26,27] In addition, both the alveolar–arterial oxygen difference ($A\text{-}aD_{O_2}$) and the dead space/tidal volume ratio (V_D/V_T) are elevated.[27] Hypercapnia is seen in idiopathic scoliosis with increasing age or in scoliosis with coexistent obstructive pulmonary disease. Therefore, the primary abnormality in gas exchange is ventilation/blood-flow maldistribution. However, the slope of the ventilatory response to CO_2 in scoliosis is less than the normal range. The dominant determinants of the slopes of the minute-volume and tidal-volume responses to CO_2 are the mechanical properties of the respiratory system.[24]

Therefore, in idiopathic scoliosis, the primary abnormalities caused by the deformity are reduced lung volumes, reduced compliance of the chest wall, ventilation/blood-flow maldistribution, and increased pulmonary vascular resistance. It is postulated that the reduced ventilatory response to CO_2 associated with severe deformity impairs the ventilatory compensation for ventilation/blood-flow maldistribution and therefore contributes to hypercapnia and hypoxemia. Superimposed on these abnormalities is age-associated deterioration in the arterial blood gases which is thought to result from deterioration of the mechanical properties of the lungs.[28] The ensuing increased ventilation/blood-flow maldistribution increases the ventilatory requirements, and hypercapnia is a manifestation of the failure to meet these requirements. Respiratory failure is thus the culmination of a spectrum of functional abnormalities caused by the effects of deformity on all the major components of respiratory function and the interaction with age-associated impairment of respiratory function (fig. 2).

Scoliosis Associated with Neuromuscular Disease

A wide variety of neurologic and neuromuscular diseases is associated with scoliosis (table 1). No attempt has been made to provide an encyclopedic description; rather, the clinical and neurologic features of various examples are discussed.

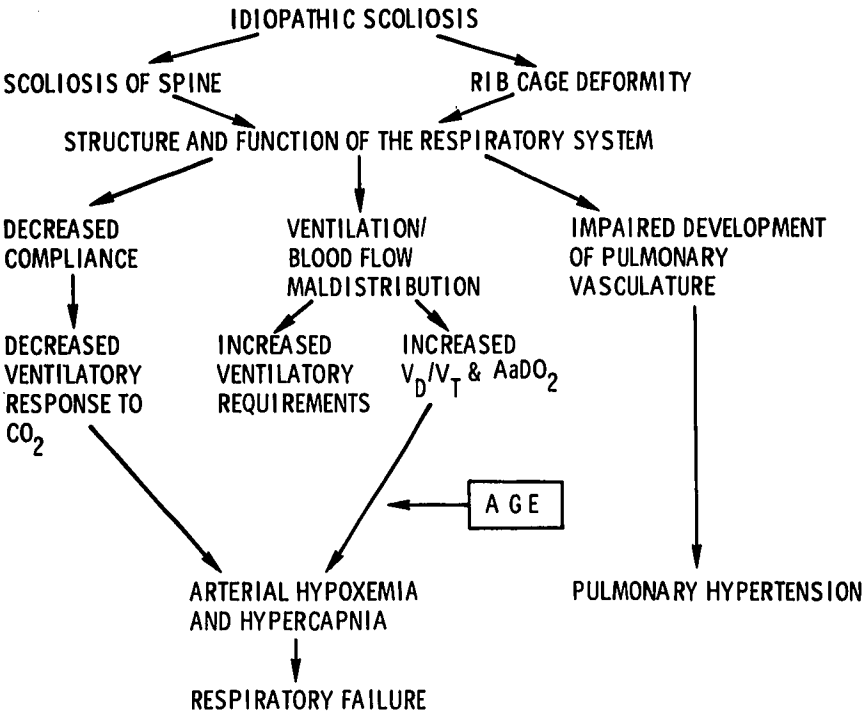

FIG. 2. The factors in idiopathic scoliosis that contribute to respiratory function abnormalities and failure. From Kafer.[15]

POLIOMYELITIS

Scoliosis following poliomyelitis is caused by an imbalance of trunk muscles during growth and is usually seen within two years following acute poliomyelitis. The important factors that determine the prognosis of the scoliosis are the muscle function, the severity of muscle imbalance, the site of the curve, and the age at which acute poliomyelitis occurred. As a result of a greater rate of progression of the curve after cessation of growth than in idiopathic scoliosis, the prognosis of paralytic scoliosis (poliomyelitis) is worse than that associated with a comparable idiopathic curve. Collapse of the rib cage is often a feature, the ribs lying almost vertically, and there may be little or no movement of the ribs during breathing.

SYRINGOMYELIA

The clinical triad on which a diagnosis of syringomyelia is based is 1) dissociated sensory loss over neck, shoulders, and arms (loss of pain and temperature with preservation of sense of touch); 2) amyotrophy; 3) thoracic scoliosis. The condition is caused by an abnormal cavity that occupies the central parts of the spinal cord in the cervical region but which may extend up into the medulla oblongata or downward into the thoracic segments. The cavity progressively replaces the gray matter of the posterior and anterior horns of the spinal cord and interrupts the crossing pain and temperature fibers in the anterior commissure. The cause is unknown, and a familial incidence is rare. Symptoms begin in late childhood, adolescence or adult life and progress irregularly, often being arrested for long periods of time. Scoliosis may appear before other evidence of the disease.

FRIEDREICH'S ATAXIA

This is the classic form of hereditary ataxia, and it forms a relatively distinct symptom complex. The disease generally runs true to form, although it overlaps other heredodegenerative syndromes, particularly the chronic familial polyneuropathies and progressive optic atrophy. The clinical features result from involvement of the spinocerebellar pathways (ataxia, intention tremor, and scanning speech), pyramidal tract degeneration (extensor plantar reflexes), peripheral neuropathy (absent tendon reflexes), impairment of position and vibration sense, and, in some patients, other sensory loss and optic atrophy. In addition to scoliosis, there is a peculiar foreshortening and high arching of the feet (pes cavus). The progressive muscle weakness involves the respiratory muscles and leads to respiratory failure. Survival beyond early adult life is rare.

NEUROFIBROMATOSIS (VON RECKLINGHAUSEN'S DISEASE)

The features of neurofibromatosis are irregular, pigmented areas (*café-au-lait*) in the skin most promi-

nent over the trunk and numerous neurofibromas. Most of the neurofibromas are asymptomatic; however, in certain locations they create abnormalities in neural function by compressing adjacent structures. These sites include the intervertebral canal, where the fibromas compress the spinal roots, and the cerebellopontine angle, where the tumor compresses the acoustic nerve and the medulla oblongata. In addition, there is a large spectrum of nervous-system and skeletal abnormalities described to be associated with von Recklinghausen's disease.

Muscular Dystrophy

There is substantial variation in the skeletal deformity secondary to muscle weakness due to muscular dystrophy. The best-described example is Duchenne muscular dystrophy. The severe sex-linked recessive form is usually manifest in the second or third year of life, and therefore involves the beginning stages of locomotion. There is an early involvement of the pelvic–femoral muscles, causing an awkward gait and inability to run. Later, the shoulder girdle and the trunk muscles are involved, and usually, by the age of 10 years, the child is confined to a bed or wheelchair. Cardiac involvement is usually observed late in the disease, and the abnormalities include tachycardia, prolongation of the P–R interval, bundle-branch block, and S–T segment elevation or depression. Death usually occurs during the second decade of life from respiratory and/or cardiac failure associated with spinal deformity and obesity.[29,30] Wilkins and Gibson[31] classified the types of spinal deformity, and thereby provided a basis for an orthopedic approach to management. They proposed that early support of the spine prevents the development of kyphosis and ameliorates the rapid progression to wheelchair, bed, and respiratory and cardiac failure. Inkley and associates[29] argue that pulmonary function studies, which include arterial blood gas determinations, lung volumes and maximal inspiratory and expiratory pressures,[32] contribute to clinical care by facilitating prognosis and allowing determination of the appropriate intensity of acute care in patients in whom pulmonary infection or respiratory failure develop. Based on the clinical experience of Inkley *et al.*, significant hypercapnia in the absence of infection is indicative of a poor prognosis. When infection was present, however, the patient often returned to his preinfection status after appropriate therapy.

Respiratory Function

In order to provide an overall view of the effects of scoliosis associated with neuromuscular disease, the respiratory function abnormalities in this group are described together.

The respiratory function abnormalities are the result of a number of mechanisms. These include the skeletal abnormalities of scoliosis, abnormalities in the central respiratory control system and in the supraspinal innervation of the motor neurons of the respiratory muscles, and the loss of muscle function due to lesions of the motor neurons and peripheral nerves or myopathy. The effects of respiratory muscle-function impairment are dependent on both the distribution (inspiratory *vs.* expiratory muscles) and the severity of loss of muscle function. The respiratory function abnormalities may be further complicated by impairment of the defense mechanisms of the airways caused by loss of control of the larynx and pharynx, ineffective cough, and reduced or infrequent large breaths. In general, the prognosis of scoliosis caused by neuromuscular disease is worse than that of idiopathic scoliosis, and, over a period of years, the condition progresses to irreversible respiratory failure.

The effects of respiratory muscle weakness, its distribution and secondary changes in lung mechanics are well illustrated by the changes documented in poliomyelitis. In ambulant patients with poliomyelitis, the inspiratory capacity and VC are reduced as a result of the loss of inspiratory muscle force. The loss of expiratory muscle force also reduces the expiratory reserve volume and increases the RV.[33–36] The FRC and the compliance of the lung and the total respiratory system are often normal. In patients with severe poliomyelitis and in patients receiving long-term ventilatory assistance, the reductions in VC and TLC are extreme, and FRC, the compliance of the lung, and the total respiratory system are also reduced.[35,37–40] The reduced compliance of the lung and reduced FRC are caused by reduced pulmonary expansion and its effect on the elastic properties of the lung and atelectasis.[41] It has also been demonstrated that although bilateral diaphragmatic paralysis is associated with a wide range of reductions in VC (30–65 per cent predicted), characteristically when the patient changes posture from a standing or seated position to a supine position, there is a further reduction in VC by about half.[42]

The abnormalities in gas exchange are due to ventilation/blood-flow maldistribution as evidenced by increased A-aD_{O_2} and V_D/V_T with reduced Pa_{O_2} and normocapnia in ambulant patients.[36] However, when associated with impaired automatic control of ventilation and/or marked inspiratory muscle weakness, hypoventilation with hypercapnia and hypoxemia is a major contributor to respiratory failure. Brain-stem

abnormality[43,44] as well as hypercapnia and impaired ventilatory response to CO_2 greater than would be anticipated from the reduction of VC,[44–46] indicate impairment of the automatic regulation of ventilation. Where the automatic control is impaired, there is extreme sensitivity to central nervous system depressant's as revealed by respiratory depression from narcotics and general anesthetics.

Orthopedic Management of Scoliosis

The Milwaukee brace is the most significant contribution to the nonsurgical management of idiopathic scoliosis.[11,14,47,48] The early application of this brace to a moderate curve may result in the correction of the curve. When the brace is maintained until full skeletal maturity, the correction is stable when the brace is removed. There are no data of the effects of the Milwaukee brace on respiratory or cardiovascular function.

Spinal fusion has a long history and recently has been combined with mechanical reduction of the scoliosis.[49,50] Harrington's technique, which uses posteriorly placed distraction bars, has provided a practical and relatively safe method of obtaining maximal correction of many curves. Several anterior approaches, including that of Dwyer, have been developed.[51,52] Although correction decreases the angle, improves the cosmetic features, may relieve or prevent back pain, and prevents progression, the effects of operative procedures on the long-term prospects of respiratory and cardiovascular functions are unknown. The majority of short-term studies have shown that there was either no significant increase, or even a decrease, in the vital capacity following corrective surgical intervention.[53–56]

Most patients in whom posterior fusion with instrumentation is performed have idiopathic scoliosis or scoliosis caused by poliomyelitis. Because of the progressive nature of the deterioration of muscle function in scoliosis associated with degenerative neuromuscular disease, few of these patients are candidates for corrective orthopedic surgical operations.

Anesthetic Management of Scoliosis

Preoperative Assessment and Anesthetic Management

Preoperative clinical evaluation for patients with scoliosis should focus on the patient's respiratory and cardiovascular status; his or her history of respiratory failure; possible coexistent respiratory or cardiovascular disease; and the type, status, and prognosis of any associated neuromuscular disease. In addition, a quantitative estimate of severity of the scoliosis in terms of the angle of scoliosis is desirable. The possible loss of expiratory abdominal muscle function (not uncommon in poliomyelitis, other neurologic diseases, and in myopathy) with resulting impairment of cough, a major defense mechanism, should be evaluated.

The basic respiratory and cardiovascular investigations should include measurement of the VC (seated and supine), forced expiratory volume and its derivatives, together with acute response to bronchodilator aerosol, resting arterial blood gases, and an electrocardiogram. In spite of the complexity of the functional abnormalities in scoliosis resulting from neuromuscular disease, the laboratory assessment is fundamentally the same as for any other respiratory disease. Although it may be argued, on the basis of Black and Hyatt's study,[32] that the maximal static expiratory and inspiratory airway pressures are a more sensitive indication of loss of muscle power, the measurement of the vital capacity is available to all clinicians. When vital capacity is evaluated with respect to the total spectrum of possible functional abnormalities, it will provide practical clinical guidance for management. The vital capacity, however, reflects a voluntary maneuver and is not a guide to the function of the automatic respiratory control system.

The choice's of premedicant and anesthetic technique (general or regional) are determined by the type of operative procedure, the patient's repiratory and cardiovascular status, the presence of anatomic abnormalities, and associated neuromuscular disease. Young patients with idiopathic scoliosis (with the exception of progressive infantile scoliosis), as well as the majority of patients with scoliosis due to poliomyelitis, tolerate anesthesia and surgical procedures without difficulty. The problems encountered occur in patients with respiratory insufficiency and cardiovascular abnormalities due to idiopathic scoliosis or scoliosis-associated neuromuscular disease. These problems include respiratory failure caused by anesthetic drugs or the effects of operative procedures, cardiac failure resulting from the difficulty in estimating fluid requirements and blood replacement, and cardiac failure secondary to respiratory failure.

In the presence of impaired respiratory function with hypercapnia, the best choice if appropriate to the type of surgery is regional anesthesia, unless there is a possibility of further impairment due to complications such as phrenic nerve blockade, pneumothorax, or a high subarachnoid blockade. Theoretically, because of the early degenerative changes and anatomic abnormalities, epidural and subarachnoid blocks may be technically difficult to attain, and they may result in incomplete or patchy blockade. To date, the only

data suggesting a high success rate of subarachnoid blockade are anecdotal. When the operation takes several hours, a subarachnoid block (or epidural) does not provide relief from discomfort, and, therefore, the anesthetic is often supplemented by narcotics. An alternative technique, used in pelvic surgery and hip replacement, is to combine a subarachnoid block with light general anesthesia (nitrous oxide), intubation, controlled or assisted ventilation, and sufficient narcotic or inhalational anesthetic to ensure patient comfort and amnesia. Where general anesthesia is required, a technique similar to that used for corrective orthopedic surgery is acceptable, such as a narcotic-relaxant combination with nitrous oxide and oxygen and controlled ventilation. Controlled ventilation is continued until the patient has recovered from the effects of anesthetic drugs and the requirement for analgesia has diminished.

There are however five other potential anesthetic problems in the management of a patient with scoliosis and the associated neuromuscular diseases. These are malignant hyperthermia, sustained skeletal muscle contractions, excessive potassium release, myoglobinuria, and cardiac arrhythmias.

Malignant Hyperthermia

In Britt and Kalow's[57] statistical review of malignant hyperthermia, six of the 89 patients with an episode of malignant hyperthermia with rigidity had idiopathic scoliosis. Relton and associates describe in detail three of these patients, all of whom received succinylcholine, which resulted in hypertonicity.[58–60] In one operation, anesthesia was discontinued, and the operation was deferred until a later date, at which time anesthesia was successfully managed with nitrous oxide and a narcotic analgesic.[59] However, in the other two operations, anesthesia was continued with methoxyflurane. Both of the patients died.[58,60]

Malignant hyperthermia is a rare pharmacogenetic myopathy in humans, which in many instances is inherited as an autosomal dominant with one or more weak recessive modifying genes.[61] Affected patients are susceptible to acute hyperthermia, which may be triggered by a variety of drugs, including potent inhalational anesthetics and muscle relaxants, particularly succinylcholine and amide-type local anesthetics.[62] The incidence is 1/15,000 anesthetized children and 1/50,000 anesthetized adults. Unfortunately, it is difficult to predict susceptibility to malignant hyperthermia, because the family history may not be positive and the patient may have had a previous administration of anesthesia without complications. Although the finding of an elevated serum creatine phosphokinase level may reveal a subclinical myopathy, this finding is not a reliable predicting factor for malignant hyperthermia.[63] Similarly, although caffeine and halothane induce abnormal contraction of skeletal muscle obtained in biopsy from patients with malignant hyperthermia, the ranges of response in patients with normal and malignant hyperthermia are wide and may overlap. Therefore, there are no infallible clinical data or laboratory tests available to predict susceptibility to malignant hyperthermia.

It is, therefore, critically important to be alert for possible early evidence of malignant hyperthermia. This evidence includes the absence of relaxation or hypertonicity in response to succinylcholine, a rise in body temperature, an elevated heart rate or ventricular arrhythmias,[64] and hypercapnia. The key to successful management of malignant hyperthermia is the early cessation of administration of potent inhalational anesthetics and muscle relaxants; cooling; supportive respiratory, cardiovascular and acid–base procedures; and administration of drugs such as dantrolene which lower free ionized intracellular calcium.

A patient known or suspected to have malignant hyperthermia should not be denied anesthesia for a surgical operation. As previously described, even anesthesia for major surgical procedures can be managed with nitrous oxide and narcotics.[59,61]

In reviewing the published reports of myopathies associated with malignant hyperthermia, the major groups described are those by Denborough and associates[65] and Isaacs and Barlow.[66] They reported families with a subclinical myopathy that also had a high incidence of elevated creatine phosphokinase. Rarer clinical myopathies associated with malignant hyperthermia include central core disease,[67–69] muscle adenylate kinase deficiency,[70] Schwartz-Jampel syndrome,[71,72] and a syndrome described by King and Denborough.[73] Some of these syndromes have skeletal abnormalities, including scoliosis and kyphosis.

Sustained Skeletal Muscle Contraction

The second potential anesthetic problem is the sustained contraction of skeletal muscle in response to succinylcholine in patients with myotonic muscular dystrophy. The sustained contraction may result in difficulty in ventilation and may also prevent intubation. Succinylcholine should therefore be avoided in patients with myotonic dystrophy.

Excessive Potassium Release

A potential problem is a rise in serum potassium following the administration of succinylcholine in the presence of neurologic motor deficits and the ensuing

cardiac arrhythmias. Gronert and Theye[74] consider succinylcholine contraindicated for patients with neurologic disorders involving motor deficits; however, it is not documented whether a hyperkalemic response to succinylcholine is a problem in chronic heredodegenerative disorders associated with scoliosis. Although hyperkalemia has been reported to occur following succinylcholine administration in patients with muscular dystrophy, the hyperkalemic response is not usually of great magnitude and is not consistent.[75,76]

Myoglobinuria

A fourth potential problem, myoglobinuria, may lead to acute renal tubular necrosis and renal failure. Myoglobinuria is a manifestation of muscle trauma and is also seen after patients with certain myopathies have exercised. These myopathies include McCardle's syndrome and muscle carnitine palmityltransferase deficiency. Moore and associates described massive myoglobinuria following the administration of succinylcholine and halothane in patients with nonprogressive familial muscular dystrophy.[77] The patient and several family members had elevated serum creatine phosphokinase of isoenzyme type III, which is characteristic of skeletal muscles.

Cardiac Arrhythmias

Cardiac arrhythmias, the fifth potential hazard, are a well-documented complication of anesthesia in muscular dystrophy, particularly Duchenne's muscular dystrophy.[78–80] Arrhythmias are evidence of myocardial involvement in muscular dystrophy, and they may be precipitated by potent inhalational anesthetics, such as halothane, and succinylcholine. They may also be caused by hyperkalemia, excessive sympathetic or vagal tone, and hypercapnia.[78]

Intraoperative Monitoring

In addition to routine intraoperative monitoring, which includes body temperature measurement and the electrocardiography, intraarterial blood pressure and blood-gas monitoring is indicated for corrective orthopedic surgery or any other major surgical procedures in patients with scoliosis. Where there is electrocardiographic evidence of right ventricular hypertrophy, or evidence of pulmonary hypertension, severe blood-gas abnormalities, or coexistent cardiovascular or pulmonary disease, a flow-directed pulmonary arterial catheter with thermodilution capability should be inserted. Measurement of the pulmonary arterial and pulmonary capillary wedge pressures, and measurement of cardiac output, will assist in the fluid and pharmacologic management of a patient having a major surgical procedure. There are no known specific anatomic abnormalities in patients with scoliosis (except for the association with congenital heart disease) that would make insertion of a flow-directed pulmonary catheter difficult. In order to avoid the complication of pneumothorax, the internal jugular-vein site of insertion is preferred to the subclavian-vein site. Precautions recommended for avoiding complications in patients with pulmonary hypertension should be followed.[81] These include: continuous observation of pulmonary arterial pressure waveform during balloon inflation and discontinuance of inflation when pulmonary wedge-pressure is achieved; inflation of balloon to wedge pressure for no more than two respiratory cycles, or for no longer than 10 to 15 sec;† and continuous monitoring of pulmonary arterial waveform to prevent accidental prolonged wedging of the catheter.

Orthopedic Surgery

The anesthetic problems encountered in orthopedic surgery for scoliosis include: management of a patient in the prone position; replacement of lost blood (which may be extensive); complications of hypotensive anesthesia when used to control blood loss; hypothermia due to heat loss from the extensive exposed area, aggravated by a cool operating room and a failure to administer warm blood and fluids; and hemopneumothorax caused by tearing of the pleura during surgical manipulation.[82–88] Recently, in order to prevent paraplegia caused by ischemia of the cord, attention has been focused on methods to evaluate the function of the spinal cord during surgical procedures.

A narcotic–nitrous oxide–relaxant technique with controlled ventilation, supplemented as necessary by inhalational anesthetics, such as enflurane or halothane, is the most common technique used for the management of patients during corrective orthopedic surgical procedures for scoliosis. The possible advantage of hypotensive anesthesia used to control blood loss in scoliosis is offset by the problem of hypotension causing spinal-cord ischemia.[89] The majority of young patients who have orthopedic surgical procedures are able to breathe spontaneously on immediate recovery from anesthesia and operation. However, in the presence of marked preoperative abnormalities in respiratory function, such as hypercapnia, or complications of anesthesia or surgical procedures, it may be necessary to control ventilation until recovery.[88]

The incidence of paraplegia following operations

† Edwards Laboratory, California. Product Bulletin—Swan-Ganz Flow Directed Catheters, 1979.

for treatment of scoliosis varies between series and is thought to occur more often when the deformity is severe, congenital, or associated with kyphosis. The total incidence of neural problems in a series compiled by the Scoliosis Research Society in its Morbidity Reports for 1975 and 1976 was 1.17 per cent (114 of 9,680 patients); in 54 patients (0.56 per cent), there was partial or complete paraplegia.[90] It is postulated that traction on the spinal cord caused by the correction of the curve causes occlusion of the arterial blood supply of the cord. Detecting the compromised cord function during the operation is essential, and two methods to accomplish this are currently being investigated. Hardy and associates[89] describe the use of evoked cortical responses of somatosensory potentials. Vauzelle and associates[91] monitor cord function by waking the patient up after full spinal distraction with Harrington instrumentation and then testing the motor function of the lower limb. This method is now gaining growing acceptance by orthopedic surgeons.[90] After full distraction, neuromuscular blockade is reversed with atropine and neostigmine. If necessary, a narcotic antagonist is administered to achieve spontaneous ventilation and response to vocal commands but care should be taken that the patient does not begin to struggle. When spontaneous ventilation is regained, patients are ventilated with 100 per cent oxygen. The patients are first asked to squeeze their hands and then to move their feet. If there is absence of movement or weakness of one or both feet, but the patients can move their hands, then the distraction is decreased and the test is repeated. On completion of testing, the patient is given diazepam and the anesthesia continued as before. The patient is informed of the procedure before the operation to avoid causing psychological trauma. The test is not performed if the patient is emotionally unstable or is mentally defective.[90]

During the last decade, orthopedic surgeons, together with thoracic surgeons, have made increasing use of the anterior approach to spinal surgery, including surgery for scoliosis and kyphosis.[51,52] Surgery for scoliosis that involves a transthoracic approach with detachment of one dome of the diaphragm may be associated with a greater degree of immediate postoperative respiratory insufficiency than the spinal fusion procedure with Harrington instrumentation. Rib resection has also been used to control the progression of scoliosis and is still used in some centers, particularly for the management of infantile idiopathic scoliosis.[92,93]

Thoracic Surgery

Although two deaths occurred in a series of ten patients having thoracotomy on the side of the concavity of the curve, and no death occurred in a series of 15 patients in whom thoracotomy was on the side of the convexity,[94] the results of most regional lung function studies have shown that the side of the *convexity* makes a greater contribution to gas exchange.[95,96] Shannon and associates[97] state that the lung on the side of the concavity is compressed. These data suggest that pneumonectomy on the convex side may leave insufficient lung for gas exchange. It should be noted, however, that the regional lung function data are inconsistent. Neither Bake and associates[98] nor Dollery and associates[99] were able to show consistent differences in lung function between the convex and concave sides.

The interpretation of the significance of the differences in lung volume and gas exchange between the hemithoraces may be further complicated by regional abnormalities in the mechanical properties of the lung and chest wall, and by regional differences in inspiratory muscle function (diaphragm particularly) as in poliomyelitis and other forms of scoliosis associated with neuromuscular disease. Therefore, each patient should be individually evaluated for thoracic surgery with pulmonary function on each hemithorax, and, if necessary, right heart catheterization.

Management of Pregnancy and Delivery

There are several case reports of pulmonary edema following delivery in patients with severe scoliosis.[100,101] However, in spite of these reports of complications following delivery, the data are as yet anecdotal, physiological studies of the effects of reduction in vascular capacity and redistribution of blood volume following delivery have not been done. To what extent poor management which might have included excess fluid or blood administration and/or excess narcotics contributed to the reported complications is impossible to assess retrospectively. Based upon available pathophysiologic data, it may be predicted that the more serious problems will occur in a patient who is in the 35–45 year age group with severe scoliosis, or in a patient with scoliosis associated with severe neuromuscular disease, and particularly in a primipara experiencing prolonged labor in whom fatigue develops.

Summary

Scoliosis, of which the most frequent and important entity is idiopathic scoliosis, leads to respiratory and cardiovascular function impairment, which is characterized by reduced lung volumes, ventilation/blood-flow maldistribution, impaired chemical regulation of ventilation, and increased pulmonary vascular resistance. Superimposed on these abnormalities are age-

associated deterioration in respiratory control and mechanical properties of the lungs and/or chest wall with a reduction in alveolar ventilation and a decrease in Pa_{O_2} and an increase in Pa_{CO_2}. There is a complex pattern of respiratory function abnormalities in scoliosis associated with neuromuscular disease, which often results in greater impairment of ventilatory control and gas exchange and a more rapid progression to respiratory and cardiac failure. In addition, pulmonary function may be further impaired by inadequate defense of the upper airway and inadequate cough.

Anesthesia is most often needed for corrective orthopedic surgery. Since this is usually performed in a young patient with idiopathic scoliosis, the management of anesthesia is usually uncomplicated. Nevertheless, as with any patient with respiratory impairment, objective preoperative evaluation is necessary, and this should include measurements of the vital capacity and forced expiratory volume with its derivatives, analysis of arterial blood gases, and an electrocardiogram. A narcotic–relaxant technique with controlled ventilation is commonly used. The temperature, electrocardiogram, urine flow, and central venous pressure, intraarterial blood pressure, and blood gases should all be monitored continuously. To prevent neurologic defects caused by compromised spinal cord arterial blood supply, there is growing acceptance of the practice of awakening the patient after distraction of the curve and testing the patient's lower-limb function.

Management of anesthesia, in the presence of more complex respiratory function abnormalities such as those associated with neuromuscular disease, or when the patient is older and has scoliosis or coexistent systemic disease, requires making the choice of regional *vs.* general anesthesia or deciding on combination of regional and light general anesthesia. It must also be determined whether it is necessary to monitor pulmonary arterial and wedge pressures and to measure cardiac output with a flow-directed pulmonary arterial catheter with thermodilution capability. Planned postoperative controlled ventilation until recovery from the effects of general anesthetics, narcotic analgesics, and muscle relaxants may be indicated. As described in the text, some procedures, including thoracic resection and obstetrical delivery, require individual evaluation of both the patient and the planned procedure.

In addition to the complications of respiratory and/or cardiac failure due to problems in the management of anesthesia in patients with severe scoliosis or scoliosis associated with neuromuscular disease, anesthesia has also been associated with malignant hyperthermia, abnormal skeletal muscle responses to depolarizing muscle relaxants, hyperkalemia, myoglobinuria, and cardiac arrhythmias. Although these are rare complications, most are predictably associated with specific types of muscular dystrophies or myopathies that can be diagnosed preoperatively. However, malignant hyperthermia cannot be reliably predicted from clinical or laboratory data except for certain specific associated myopathies. Therefore, early detection and appropriate therapy are critically important to successful management during anesthesia.

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