Current Practice

RESPIRATORY TRACT DISEASE

Chest Deformities

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Deformity of the chest may arise from maldevelopment of the ribs, the sternum, or the vertebral column; or from an abnormality in the lung, such as a large lung cyst. In each case the deformity may be congenital in the sense that the defect is present at birth, or it may be acquired.

Deformities of Ribs

Congenital deformity of the ribs may occur as a cervical rib, or as a bifid anterior end to a rib. The latter, though regarded as abnormal in man, is in fact normal development in some other mammals. Less commonly there may be complete or partial fusion of two ribs. Rarely there may be complete absence of one or more ribs. These rib disorders seldom of themselves produce much alteration in the contours of the thorax, though a rib which is longer or shorter than normal may do so; they are often detected only by chest radiography.

Deformity of Sternum

The sternum may be congenitally malformed with a central prominence, but a more common deformity is that of sternal depression. This arises either from maldevelopment of the parts of the sternum or from an excessive pull of congenitally hypertrophied anterior leaves of the diaphragm. Occasionally some sternal prominence occurs in late childhood for no obvious reason. More commonly it occurs as a secondary phenomenon, the primary cause being, for example, Pott's disease of the spine, in which the collapsed and wedged vertebrae lead to an angular kyphosis and a great increase in the antero-posterior chest diameter. Sternal prominence may also be found in patients with bronchial asthma developing in childhood. Cinematography of a young child's chest during an acute asthmatic attack demonstrates well the way this deformity is produced—as the diaphragm on inspiration draws air past the airway obstruction, its inward pull from the lateral attachments thrusts the sternum forward.

Abnormally formed ribs seldom require treatment unless they produce symptoms by pressure on blood-vessels or on the brachial plexus at the thoracic inlet. Similarly, sternal prominence is not usually treated surgically, prevention being the major aim. The threat of permanent deformity occasionally provides an indication in severe childhood asthma for long-term treatment with corticosteroid drugs. Occasionally, if treated in this way and in good time, the prominence of an asthmatic child's sternum may be seen to regress as the child grows though treatment must be continued over several years.

Sternal depression produces symptoms only if it is severe, which is a rare occurrence. In a few cases venous return to the heart may be impeded by obstructing flow through the great

vessels into the right side of the heart; surgical correction may then be justified. In the majority of patients, however, no organic symptoms arise and recent published series have lent little support to the view that surgical correction improves or prevents respiratory symptoms. This is not to say that surgery has no place in the treatment of sternal depression. It is an important cosmetic measure. In young girls moderate sternal depression often becomes unnoticeable as breast development takes place, but adolescent boys may become painfully self-conscious and surgical correction of their deformity may enable self-confidence to be regained. These cosmetic problems are best referred to a thoracic surgeon.

Ankylosing Spondylitis

To the foregoing abnormalities of shape of the chest wall should be added those resulting from ankylosing spondylitis. Here the restriction of chest expansion is usually accompanied by a relatively immobile spine and a head held in a forward-leaning position. The spine is not truly kyphotic, for there is not usually an excessive backward curve of an otherwise upright vertebral column. The general posture in ankylosing spondylitis is easily recognized and is well known. Less well known is the good respiratory performance achieved by many such patients, especially when, as usually happens, good diaphragmatic movement is present and the transfer of oxygen through the lungs into the pulmonary circulation is normal. The loss of lung volume and ventilation, which is seldom severe, is then of less importance than might be supposed, and the prognosis is accordingly good. In an analysis of 70 deaths occurring in a Ministry of Pensions series of patients with ankylosing spondylitis only five died from acute respiratory disorders.

Scoliosis

In the deformities so far mentioned the vertebral column is straight or may be kyphotic. But perhaps the most important group, certainly those requiring greatest care in management, include those with lateral spinal curvature (scoliosis). In these the associated chest deformity may become very severe and cardio-respiratory function may be disturbed.

Scoliosis may be postural or structural. The former, often found in adolescence, can be corrected by voluntary effort, and disappears if the spine is flexed. This type is physiologically unimportant. Structural scoliosis is always a matter for care and in childhood ought always to be supervised by an orthopaedic specialist, whether or not any physician is involved in the care of the patient. Scoliosis may arise from developmental abnormalities of the vertebral column, such as hemi-vertebra. It is then correctly called congenital scoliosis, even though scoliosis may not develop until later on. It may also arise from local causes in association with generalized developmental

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disorders, of which neurofibromatosis, Friedreich's ataxia, muscular dystrophy, and Marfan's disease are among the more important. Though such diseases are of congenital origin the cause of the spinal curvature is not known, and is hence called "idiopathic."

Idiopathic scoliosis may more frequently be found without any congenital or other disease being present, and indeed this is the largest group. It may start in early middle or late childhood, and is by convention called infantile if of onset between 1 and 4 years, juvenile if between 5 and 9 years, and adolescent if at 10 years or beyond. Like other forms of scoliosis, it tends to worsen most quickly when spinal growth is most rapid, and it rarely changes after bony growth has ceased. Indeed, many orthopaedic centres discharge scoliotic patients at this time.

The only established facts regarding the aetiology of adolescent idiopathic scoliosis are that it is four times as common in girls as in boys, and in four out of five patients the spine curves convexly to the right. Little else is known of its aetiology. The clinical appearance of an otherwise healthy person with unexplained spinal curvature is so characteristic that the term "idiopathic" tends to be used to describe this type of scoliosis as if it were a single entity. This may be misleading aetiological. Unless the true meaning of the word is remembered one may miss other features such as might suggest a congenital cause. In contrast with idiopathic scoliosis, wherein the musculature is normal, is the group with paralytic scoliosis—that is, spinal curvature following poliomyelitis in which the shoulder girdle and erector spinae are affected. Severe deformity of the spine and chest may occur, usually commencing about a year after the onset of the infection.

Prognosis

These three types of scoliosis, congenital, paralytic, and the miscellaneous idiopathic group, have been studied closely by orthopaedic surgeons, especially over the last 20 years. They are now able to foresee with fair accuracy the likely outcome of a particular case. The prognosis is based on the type of scoliosis, the site of the curve, and the age of onset. Generally speaking, prognosis for high thoracic curvature is worse than for that lower down. If the expected prognosis is good it may be decided merely to observe the patient, and to make sure that a mild degree of curvature does not worsen. If the curvature is likely to increase, a spinal support, such as the Milwaukee brace, may be used. This extends the spine by pushing the head upwards from a corset anchoring the brace round the hips. At the Calot Institute in France a successful treatment has been to use a long-term plaster-of-Paris jacket, renewed at intervals over a period of years, and combined with vigorous physiotherapy to encourage good chest movement within the plaster. Either method, if adequate to hold the patient in a good position to the end of the growth period, may be successful in many cases. Sometimes, however, this conservative treatment may prove inadequate and may need to be followed by spinal fusion. This is usually done as an extra-thoracic procedure, in which a bone graft or bone chips are laid around the spinous processes of the vertebral column.

Lung Function

Much interest has been taken in the physiological effects of spinal curvature on patients. Some of these effects have been noted since the time of Hippocrates. Particular interest has been shown in recent years in the changes in heart and lung function associated with scoliosis. Physiological studies show that as spinal curvature increases there is a progressive fall in total lung volume and its subdivisions. The fall in vital capacity is particularly marked in the more severe grades of

paralytic scoliosis. As might be expected from the intercostal and diaphragmatic weakness often present in paralytic patients, this lowered vital capacity is matched by a reduction in ability to ventilate the lungs. The transfer of oxygen through the lungs into the blood-stream is reduced when lung volumes are small, but the mixing of air in the lungs in these patients is seldom abnormal. The absence of any airway obstruction, even though the bronchi may be distorted, is a surprising though striking feature. Lung "stiffness" is seldom seriously increased. The most important alterations in physiology are the small lung volume found in severe cases and the high consumption of oxygen due to the work of breathing. Radiographic and fluoroscopic examination of the lungs is valuable, the former showing in particular the severity of thoracic curvature and the blood vasculature of the lung fields, while the latter shows the extent of diaphragmatic movement, which is often crucial in assessing operative risk.

Heart Disease

Since scoliosis is an anatomical, not an aetiological, diagnosis, generalizations based only on the degree of scoliosis present may be inaccurate. This is true not only of respiratory function: it is also true of the heart. In several generalized developmental disorders both the heart and the vertebral column may be abnormal. Marfan's disease, Friedreich's ataxia, and the Klippel-Feil syndrome are examples of conditions in which scoliosis and abnormal cardiac states often coexist. Examination of large numbers of scoliotic children who were otherwise apparently normal has revealed three important facts. Firstly, cardiac murmurs are not caused by scoliosis, however severe. If a murmur is present it is usually due to a congenital heart lesion. Secondly, the heart is usually in a normal position within the thorax. Gross displacement seldom occurs. Thirdly, the electrocardiogram is normal when taken from the customary limb- and chest-lead positions. This knowledge enables one to detect otherwise unsuspected cardiac disease. So-called kyphoscoliotic heart disease, meaning cardiac failure secondary to severe scoliosis, does not exist in children. Abnormal cardiac findings in childhood indicate the presence of an abnormal cardiac state, quite apart from the presence of scoliosis.

In later life a proportion of severe scoliotics may develop heart failure. Of the numerous theories put forward to explain why this happens, the most convincing is the suggestion that an upset in the ratio of ventilation to perfusion of parts of the lung leads to localized anoxia and a consequent eventual rise in pulmonary artery pressure. The treatment of this type of cor pulmonale is similar to the orthodox management of congestive heart failure from other respiratory causes. Why adults in their 30's and 40's develop cardiac and respiratory failure within a few months of being relatively active remains unexplained. Not until the results of a long-term study of scoliotic children are available will it be possible to learn what proportion develop cardiac or respiratory failure or the reasons that they do so. Meanwhile there is much to be said for young persons with severe scoliosis receiving medical supervision, especially if, as growth finishes, they no longer remain under orthopaedic care.

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