Bone and Joint Diseases

Management of structural scoliosis

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Scoliosis means lateral curvature of the spine from any cause. There are three types: postural, compensatory, and structural (fig 1).

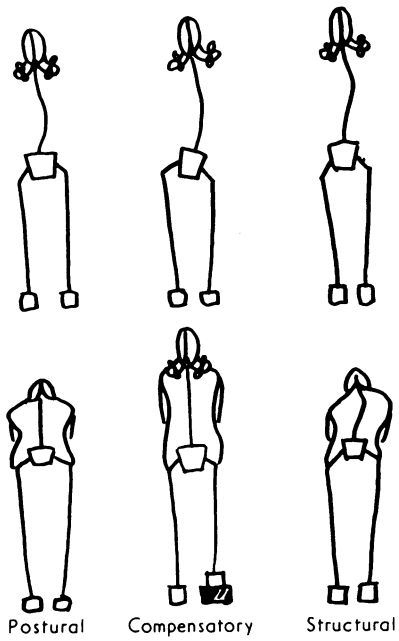

FIG 1—Classification of spinal curves according to structural changes.

Postural scoliosis is not a true deformity but is simply a habit stance that disappears with change of position; there is no anatomical abnormality and only reassurance is required.

Compensatory curve—This also is not a true spinal deformity, but an abnormal posture to compensate for a real deformity elsewhere, usually unequal leg lengths that tilt the pelvis so that the spine must curve to preserve the upright stance. The scoliosis disappears when the underlying deformity is neutralised—that is, by a raise under the short leg—though after many years fixed structural changes may develop. Treatment is directed at the primary abnormality.

Structural scoliosis, by contrast, shows anatomical changes in the spine and persists in all positions. Furthermore, the spine is rotated as well as curved, and, although we define scoliosis as lateral curvature, rotation causes much of the deformity and may appear first. The vertebrae rotate about a vertical axis so that the bodies face the convex side of the curve, and the trunk structures, rotating with the spine, become prominent on the convexity, especially in the thorax, where rib projection causes the hunch-back profile (fig 2).

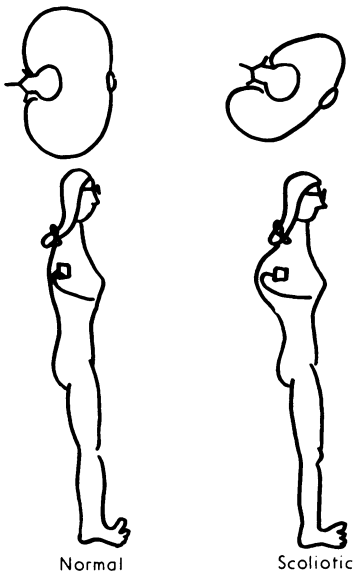

FIG 2—Rotation about vertical axis throws ribs into prominence on convexity, making silhouette kyphotic. True spinal kyphosis is usually absent and "kyphoscoliosis" is generally a misnomer.

Of these three types, postural, compensatory, and structural, only the last is primarily a spinal problem and what follows refers to this alone.

Causes

Structural scoliosis as a clinical problem starts in the growing years. It is true that adults with osteoporosis may develop minor curves, but these do not need specific treatment, and I shall consider only the common deformities of youth. About 20% are congenital, one or several veterbrae being asymmetrically malformed; neuromuscular imbalance of the trunk causes 15% (more where poliomyelitis is common) and occurs in many disorders—notably, poliomyelitis, syringomyelia, congenital cerebral palsy, peroneal muscular atrophy, Friedreich's ataxia, spina bifida, neurofibromatosis, and muscle dystrophy. Rarer causes include bone dysplasias such as osteogenesis imperfecta, while a few curves are secondary to thoracic disease or surgery. The remaining 60% of cases are of unknown origin.

Prognosis

A structural curve makes the spine permanently asymmetrical in relation to the trunk muscles and body weight, so that greater pressure is exerted on the concave side than on the

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convex. Compression inhibits growth at the cartilaginous vertebral end plates; thus increase in height of the vertebrae is retarded on the concave side and the curve increases (fig 3). For this reason structural scoliosis, however initiated, progresses throughout growth, though often at an unpredictable rate. Thereafter progression becomes much slower, but pressure absorption of disc and bone in the concavity may account for slow deterioration in adult life.

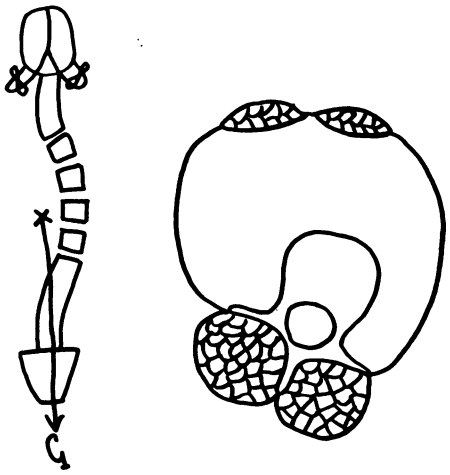

FIG 3—Lateral curve and rotation both displace vertebrae away from midline and resulting asymmetry of muscle action and body weight causes greater compression on concave side of curve.

A strange exception to this general rule of progression is a curve of unknown cause appearing in the first year of life and designated infantile idiopathic to distinguish it from the much commoner idiopathic curves of adolescent onset.[1] Most of these infantile curves disappear before 3 years of age.

Unchecked scoliosis may be extremely distressing, and (excepting the resolving infantile type) the outcome is generally worse with early onset and in the upper part of the spine. Thus, a high thoracic curve may become an obvious, even grotesque, hunchback deformity; the thoracic distortion causes cardiorespiratory insufficiency, sometimes fatal, and there is a slight risk of paraplegia from angulation of the cord. Curves at lower levels are less menacing; a lumbar scoliosis is not so conspicuous, is free of cardiorespiratory or neurological complications, but may cause severe backache from secondary osteoarthritis.

Choice of management

Treatment is arduous for both child and parents, and, though much improvement may result, the spine will not become normal. Three approaches are available: observation, external bracing, or operation.

OBSERVATION ONLY

A decision to withhold treatment does not imply that deformity is acceptable but that sometimes the medicine is worse than the disease. There are three common indications: firstly, the resolving infantile curve; secondly, a mild curve, detected when growth has ended, though it must be observed for several years to check that it is static; and thirdly, when other disabilities—for instance, severe spastic quadriplegia with mental retardation—are overwhelming. A handicapped child, however, is usually better off with a straight back than a bent one, and the mere presence of another disability is not an automatic disqualification.

BRACING

A Milwaukee brace[2] is often effective, but very restrictive. It comprises a leather or plastic pelvic girdle and a segmented collar loosely circling the neck, taking purchase on the occiput, and just touching the throat. The girdle and collar are connected by metal struts that can be lengthened as the curve improves and as the child grows, and a pad is attached to these to apply lateral pressure at the apex of the curve (fig 4). This appliance, worn day and night, often for many years, and removed only for bathing may be a daunting prospect, but on average it will arrest a mild or moderate curve. Some do better, gaining permanent improvement; others not so well and continue to deteriorate. A brace is used for: (a) a child who will need operation, but is too immature (the ideal time is just at the end of growth); (b) a slight curve in late adolescence (a brace until growth has finished may keep it minimal); and (c) a child unfit for or unwilling to have surgery.

SPINAL FUSION

Most curves need arthrodesis of all the affected segments in the straightest possible position, with internal fixation to give added security until fusion is sound. Preoperative correction is very important, and remarkable improvement often results. The technique depends on the severity and rigidity of the deformity. Moderate scoliosis is corrected a few days before operation by a plaster cast extending from the pelvis to the occiput,[3] applied with the patient on a special frame exerting both longitudinal traction and lateral pressure on the curve. More severe deformities have continuous traction for a few weeks, applied via pelvic girdle and occipitomental halter,[4] or more rigorously through a metal halo screwed to the skull[5] and traction pins through the lower femurs. The most resistant may need a halo-hoop apparatus,[6] in which threaded skewers are drilled obliquely fore-and-aft through the iliac crests so that a hoop encircling the pelvis can be attached to them, while a halo is fixed to the skull;

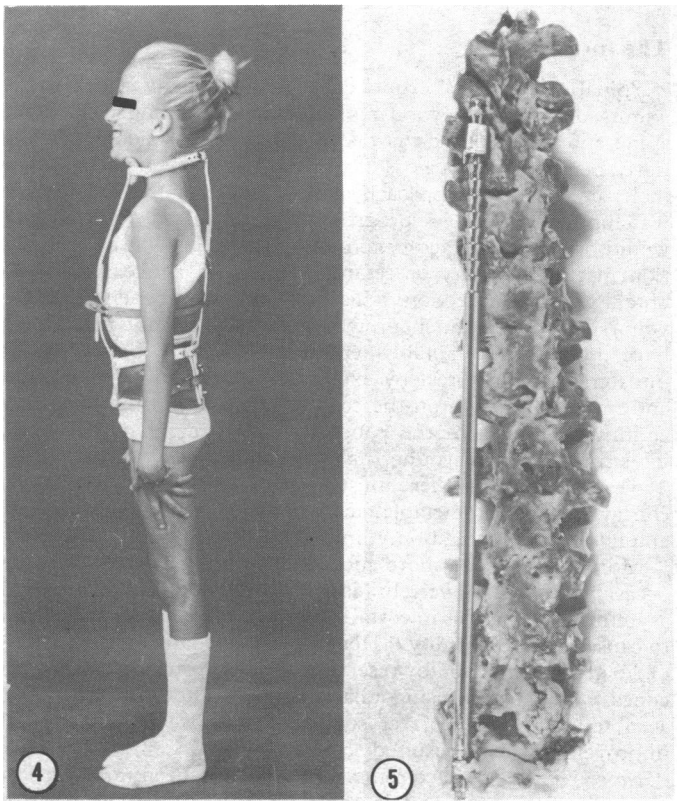

FIG 4—A Milwaukee brace, named after its city of origin. FIG 5—A spine with a Harrington rod and hooks in situ, five months after fusion.

hoop and halo are connected by four longitudinal trombone-like sliding rods that are very slowly elongated by a screw mechanism.

These drastic measures, reserved for the most severe deformities, may cause neurological damage and strict observation by an experienced team is essential.

Fusion of the whole curve follows preoperative correction. The neural arches are exposed subperiosteally, the articular surfaces of the posterior joints excised, and the resulting spaces packed with bone—either free graft from the ilium or pieces turned in from the adjacent transverse processes and neural arches. The remainder of the neural arch is then split into slivers, which are turned up and down to interweave with those from adjacent vertebrae, thus forming a continuous interlocking mass of bone fragments throughout the whole operation area, which is usually supplemented at this stage by additional free chips taken from the ilium. Finally, a Harrington distraction apparatus[7] is inserted. This comprises a hook at each end of the fusion area, the upper one implanted into the strong point of the neural arch where transverse process, lamina, and pedicle meet, the lower into a notch cut in the lamina. Between these hooks a rod is inserted, ratchet-shaped at one end to lock the hooks securely as corrective distraction is applied (fig 5). Although this implant seems most dramatic to the patient, permanent correction depends on bony fusion, not on metal, and the great value of the Harrington apparatus is that, in combination with a plaster jacket, it makes the spine stable when upright; thus as soon as the incision is healed a cast is applied and the child returns to school.[8] Plaster is usually removed six to seven months after surgery.

Posterior fusion requires good quality bone in the neural arches which sometimes—for instance, in spina bifida—is not available, and then the vertebral bodies must be fused, through a retroperitoneal or transpleural approach, or both. The discs and vertebral end plates are excised and the resulting gaps packed with bone chips; internal fixation is then applied by ring-ended screws inserted from the convex side, one across each vertebral body, the rings being joined by a longitudinal cable, which, on tightening, approximates them to correct the curve.[9] Aftercare is as for posterior fusion.

The future

Spinal fusion is a crude concept: essentially the surgeon admits that forces beyond his control are bending part of the spine and all he can do is make that part so rigid that it will never bend again. No gloss of technical refinement can hide the inelegance of this approach, but it is certainly better than watching the relentless progression of untreated deformity. It would be more satisfactory to modify spinal growth by stimulation in the concavity or retardation on the convexity and so obtain gradual correction. This would be most beneficial in the young child with much growth potential. Several procedures have been tried, notably epiphysiodesis,[10] which aims at unilateral growth arrest by excision of cartilaginous end plates and interbody fusion on the convex side only, in the hope that continued growth in the concavity will straighten the curve. This must be done young (half the adult height is reached by $2\frac{1}{2}$ years of age) and there must be good growth potential in the concavity, an unpredictable factor in scoliosis. For these reasons epiphysiodesis has had only limited success.

More subtly, if no more successfully, rib operations have an occasional use. Each vertebra is gripped by two pairs of ribs and asymmetrical changes in costal growth and mechanics are known to cause spinal deformity.[11] Hence possibly changing the pattern of rib growth or action by resection of their posterior ends on the concave side, or by fusing adjacent ribs on the convexity to act as a tether, might correct some spinal curves. Only limited improvements have resulted so far.

More recently an electronic stimulator has been implanted to produce continuous muscle contraction at a tolerable level of comfort.[12] It is hoped that such action on the convex side of the curve may cause permanent correction, but this technique is in its infancy.

Conclusion

Treatment of scoliosis is at a most interesting stage. In most cases deformity can be improved, but at a very high cost in time, inconvenience, and discomfort to the child and parents, and the established regimens are in no sense curative: some deformity always persists, and after operation some movement is lost. New approaches, however, give hope of improvements to come. Treatment is for specialised centres, but early referral will forestall severe deformity, and everyone concerned with the management of children should be watchful for spinal curvature and rotation.

I thank Mr Norman Gill for his skilled photography.

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ONE HUNDRED YEARS AGO The usual Christmas treat to the little patients in the Hospital for Sick Children, Great Ormond Street, took place last week, and a considerable number of the friends and subscribers of the charity were present on the occasion, among whom may be named the medical officers of the hospital, who always manifest great interest in the happiness of the children under their care. An immense Christmas-tree, which was literally ablaze with light and loaded with children's treasures, was speedily shorn of its beauty by the distributors, Mr W E Cant and Mr Henry Mason, the resident medical officers. The "tree" was followed by a magic lantern exhibited by Mr C Kemp Welch, one of the managers, all the slides being of an amusing character and well adapted to excite merriment. Creature comforts were not forgotten, and such small luxuries as the medical officers permitted, in the form of cake and fruit, were freely indulged in. The evening was enlivened by the singing of carols and such melodies as delight children, this part of the entertainment being admirably managed by the lady superintendent and the ladies who render invaluable voluntary help as ward-superintendents. In this hospital, all the nurses are young women (and, a noticeable fact, nearly all short of stature), and competent to amuse the children by singing, so that they were well able to "swell the full chorus" on the occasion. The enlarged hospital possesses a staff of twenty-two nurses; that is, seventeen for day attendance and five for the night duty, there being one hundred and four beds in the large wards, besides twenty beds in the special wards. The hospital has a convalescent home at Highgate, where fifty-two children are provided for. The Christmas festivities there were not on such an extended scale as at Great Ormond Street; but, under the direction of the lady superintendent (Miss Wood), and with the hearty co-operation of the medical officers (Dr Sturges and Mr Warrington Haward), the children enjoyed their afternoon extremely. A large number of friends gathered to witness the pleasure afforded to the children and to assist in the distribution of the gifts provided in abundance by thoughtful donors. In both instances, the period of amusement was not prolonged to the point of weariness, and, soon after seven o'clock, the wards of the two hospitals resumed their usual state of quietude, except that, on such a special occasion, the children were allowed to have their toy treasures in the cots until "tired Nature's sweet restorer" came to close the weary little eyes. (*British Medical Journal*, 1877.)