FUNNEL CHEST

IAN D. SUTHERLAND, SCONE, SCOTLAND

From the Eastern Regional Orthopaedic Service, Bridge of Earn Hospital

A not uncommon cause of defective posture in children is a funnel chest deformity. This is usually accepted as being incapable of correction and the child is treated by remedial exercises in the hope of improving the posture, without treating the cause. The purpose of this paper is to demonstrate the striking improvement that can be obtained by operative correction of the chest deformity.

ETIOLOGY

The first description of funnel chest in medical literature appears to have been by Bauhinus in 1596 (quoted by Brown 1940). Since then there have been a considerable number of

references, and the literature on the subject was reviewed by Ochsner and DeBaakey when they reported a case in 1939. The etiology of the condition remained uncertain until Brown (1939) suggested that the cause was a mechanical one due to a congenital shortening of the central tendon of the diaphragm. This theory does not explain the variations of chest deformity that may occur. Brodtkin (1953) pointed out that the anterior part of the diaphragm arises from the embryonic septum transversum and that failure of normal development would produce a relative weakness of this part. Contraction of the more powerful surrounding muscle will then overcome the defective part, drawing it and its lower sternal attachment backwards. Chin (1957) supported this supposition by muscle biopsies taken from the anterior diaphragm during operations for funnel chest deformity. He found marked deficiency of muscle fibres in these cases. It seems probable that this relative deficiency of the muscular element is the cause of the deformity. When the diaphragm is relaxed in expiration it assumes a dome shape. When it contracts to produce inspiration this dome tends to flatten out and increase the internal capacity of the chest. The

FIG. 1

Radiograph of a boy of fourteen years, showing gross depression of the sternum, which is only a few centimetres from the thoracic spine. Note the flattening of the chest and lower thoracic spine. (Same boy as shown in Figures 5 and 6.)

defective anterior part is overcome by the stronger musculature of the remaining diaphragm which draws the xiphoid and lower sternum towards the vertebral column. This action is seen very clearly when the patient is asked to inspire deeply. As the child grows, the sternum and xiphoid develop a fixed funnel-shaped deformity with the apex of the funnel at the sterno-xiphoid junction. In severe cases the xiphoid and lower sternum may be nearly touching the vertebral column (Fig. 1). At the same time the lower costal cartilages become drawn together and angled backwards.

The abdominal linea alba is attached at its upper end to the back of the xiphoid and continues upwards under the sternum as the retrosternal ligament. The backwards angulation of the sternum causes relative shortening of this ligament, pulling the chest down and producing an upper thoracic kyphosis.

Many patients give a familial history. Among those recorded here are two sisters, two cousins, and another child whose uncle, now an adult, is severely disabled by the condition. There seems no doubt that the deformity is inherited and occurs in certain families. It is probably much commoner than is generally thought.

FIG. 2

FIG. 3

Figure 2—Boy aged eleven years with progressive deformity. Funnel-shaped depression is accentuated by deep inspiration. This boy also shows lateral indrawing of the lower chest and flaring of the lower costal margins from pull of the defective diaphragm. Figure 3—Same boy showing effect on posture. The shoulders and upper thoracic spine and neck are pulled forwards. The lower thoracic spine is flat.

EFFECTS OF THE DEFORMITY

The deformity is often first noticed soon after birth, but in many individuals it may not become obvious until later childhood or adolescence. At this later stage alterations in the thoracic cage have occurred and the secondary changes in posture have developed. There is a characteristic depression of the sternum which is increased with inspiration. The costal cartilages attached to the lower sternum and xiphoid become angled backwards. The whole chest is flattened and the abdomen is often protuberant in contrast. There is an upper thoracic kyphosis, with the neck and shoulders pulled forward, and a flattening of the lower thoracic spine (Figs. 2 and 3).

In severe cases the thoracic viscera may be affected by the compression and distortion of the chest, and interference with respiration and cardiac function is not uncommon. Most of the patients reported here stated that they were unable to run as far as others of their age because of breathlessness, and this interfered with games and sports. The effect upon respiratory function is seldom disabling, but the effect upon cardiac function may be serious

in more severe cases. The heart is usually displaced to the left as the sternum retracts into the mediastinum. Cardiac arrhythmia, a low cardiac tolerance and systolic murmurs have all been reported (Lester 1950*a* and *b*, Ravitch 1951). Electro-cardiographic changes may be found. All the patients in this series have had electro-cardiographic examination carried out, but no abnormality of cardiac function has been noted. Nevertheless, it is interesting that all the children stated that they did not realise how much they were handicapped physically until they appreciated the improvement obtained after operation.

The deformity has a marked psychological effect. These children are usually very aware of the fact that they are different from others. They are shy, retiring and rather lonely, and take little share in the games and activities of their schoolfellows. They try to escape from such recreations as swimming and gymnastics at school, mainly because of self-conscious embarrassment rather than the physical handicap. The parents are worried about their development and find them difficult to manage at home. With the onset of physical maturity the psychological effects become more marked.

TREATMENT

A number of children with this deformity have been seen by the staff of the Children's Orthopaedic Service of this Region. Treatment was concentrated on the postural defects with the usual forms of physiotherapy. The results of such treatment in the more severe cases have been far from satisfactory. Most of them have shown progressive postural deterioration as they grew older. It was the failure to improve these children that led to operative correction of the cause.

The indications for operation are orthopaedic, physiological and psychological. The progressive postural deterioration does not respond to conservative treatment, but improvement after operation is often dramatic. Dyspnoea, diminished exercise tolerance and cardiac arrhythmia are also indications for operation, and post-operative improvement in exercise tolerance has been a feature of all our cases. Perhaps the most striking result has been the psychological change. This alone would justify the operation.

The age at which treatment is undertaken will depend upon the severity of the deformity: ideally it should take place before permanent thoracic and postural changes have occurred. At this early stage the operation is simple, but when structural changes in the chest have developed it is necessarily more extensive. The youngest child in this series was a boy aged five who was beginning to show a fixed deformity in the sternum and costal cartilages. There appears to be no reason why the operation should not be done at a younger age; but it is not always possible to predict which children will develop progressive deterioration as they grow older. By the time growth has ceased the thoracic and postural changes are permanent and full correction will not be possible even with an extensive operative procedure. The indication for operation in the late adolescent or adult will, therefore, be mainly to relieve disabling cardiac symptoms.

Technique of operation—The operation is based on the underlying pathology of a mechanical defect in the diaphragm attached to the back of the xiphoid and the adjacent sixth and seventh costal cartilages. The xiphoid and the strong retrosternal ligament must be divided from the sternum. In young children, before fixed changes have occurred or with minimal sternal deformity, this is all that is necessary.

In older children, with structural changes of the thorax, a more extensive procedure is required. Under intratracheal anaesthesia a midline incision is made from above the site of sternal angulation to below the xiphoid. The pectoral muscles are dissected from their attachments to the sternum and costal cartilages and reflected to expose the full extent of the cartilages. The recti abdominis muscles are detached from the lower cartilages. The seventh and eighth cartilages are usually crowded together, and to obtain an adequate exposure of the xiphoid they should first be resected as far down as their point of angulation. The

junction between the xiphoid and the sternum is now defined and the xiphoid is detached by a diathermy knife or a strong pair of scissors. The retrosternal ligament is firmly attached to the back of the xiphoid and is usually divided at the same time. The xiphoid retracts and the lower mediastinum and pericardium are exposed. The fourth, fifth and sixth costal cartilages on each side are resected. It is essential in doing this to remove the cartilages to beyond the point where they angle backwards. In this series the costal cartilages were removed subperichondrially. This makes the procedure more tedious, but it diminishes the risk of opening the pleura, and hardening of the chest wall occurs sooner after the operation. The backward angulation of the sternum is usually between the third and fourth cartilages. At the point where the angulation occurs, a transverse V-shaped osteotomy is performed (Fig. 4). This is easily done with a gouge. The posterior cortex is left intact. A finger is now passed behind the sternum and its posterior aspect is gently cleared of mediastinal attachments.

FIG. 4

Diagram of thoracic cage to show the detachment of xiphoid, removal of costal cartilages, and site of osteotomy in sternum.

Provided the retrosternal ligament is not reflected this procedure presents no difficulties. With the finger in the mediastinum, the sternum is elevated forwards. The posterior cortex fractures either completely or in the manner of a greenstick fracture. The wedge osteotomy is thus closed. The elevation of the sternum should be sufficient to allow over-correction because there is always a little loss of correction during healing. A strong curved needle with silk sutures is passed through the periosteum and bone of the anterior cortex to hold the sternum in its over-corrected position. Two mattress-type sutures are usually adequate. The pectoral muscles are reattached. The edges of the upper recti are sutured together to cover the mediastinum and the retracted xiphoid. The skin wound is closed with interrupted sutures. A light gauze dressing is held in place with strapping across the front of the chest.

This operation differs somewhat from those described by others in the literature. In the methods described by Brown (1939) and by Lester (1946, 1950*a* and *b*) wires are passed through the sternum and out through the skin on each side of the wound and attached to a wire ladder across the front of the chest to hold the sternum in the corrected position. Sweet (1944)

modified this procedure by stitching the divided ends of the costal cartilages to the sternum. Ravitch (1949) isolated the sternum from all its attachments and held it in position by sutures at the site of osteotomy: this would surely lead to avascular necrosis.

It seems wiser to avoid any form of external traction because the risk of infection is unjustified in an operation of this nature. It is not necessary to isolate the sternum from its lateral attachments.

Post-operative treatment—As soon as the patient recovers from the anaesthetic he is propped up on pillows and deep breathing is encouraged. It is surprising how fit these children are on the day after operation, and their subsequent progress has given no cause for anxiety. Most of them are anxious to get up on the third or fourth day. Breathing exercises are continued twice a day. The wound is healed in ten days and the patient then begins active shoulder and pectoral exercises, and a few days later is ready for more extensive postural training. The sternum and front of the chest are quite firm at the end of two weeks. From this time the patient should attend the physiotherapy department, being allowed home at the end of three weeks. Regular postural exercises must be continued indefinitely to correct the kyphosis and other postural defects, and to maintain the correction obtained.

RESULTS

This operation has been performed on twelve children varying in age from five to fourteen. In the five-year-old boy, who had early but progressive changes in the sternum and costal cartilages, the simple operation to detach the xiphoid from the sternum was all that was

FIG. 5

FIG. 6

Figure 5—Boy aged fourteen years with severe funnel chest, marked flattening of chest, and postural changes. Figure 6—Same boy six months after operation. There is still a little depression of the sternum but posture is greatly improved.

done. In the twelve months that have elapsed since operation the deformity has decreased and the child's posture and general health and development have improved greatly. The remaining eleven patients have been observed for periods ranging between six months and

FIG. 7

FIG. 8

Figure 7—Same boy as in Figure 2, showing correction of chest deformity. Figure 8—same boy as in Figure 2, showing improvement in posture after operation.

FIG. 9

FIG. 10

Figure 9—Girl aged fourteen years with chest deformity and poor posture, and marked psychological effects. Figure 10—Same girl six months after operation. Chest deformity is fully corrected and posture is normal. She showed dramatic psychological improvement.

FIG. 11

FIG. 12

Figure 11—Girl aged twelve years with progressive chest deformity and postural deterioration. Figure 12—Same girl six months after operation. Chest deformity is corrected and posture is normal.

FIG. 13

FIG. 14

Figure 13—Girl aged twelve years with severe chest deformity and postural defects. A shy, retiring child who never played with others. Figure 14—Same girl six months after operation. Chest deformity is corrected and posture is normal. Marked psychological improvement.

two and a half years. In all cases the results have been excellent. In ten the chest deformity has been corrected. In one case, that of a boy of eleven at time of operation who had severe postural and chest deformities, there is still a mild depression of the sternum, but posture, physical activity and general health are so improved that he has become a promising young athlete (Figs. 5 and 6). The other ten children in the series not only show correction of the chest deformity, but their postural and physical abilities are now those of normal children (Figs. 7 to 14). The parents of all the children and the physiotherapists responsible for their post-operative treatment have confirmed the psychological change that followed operation, which in most cases has been more dramatic than the physical improvement.

SUMMARY

1. Funnel chest deformity is a common cause of progressive postural defects in children.
2. The underlying pathology of the funnel chest is a congenital deficiency of the muscle fibres of the anterior part of the diaphragm which allows the stronger posterior element to pull backwards the xiphoid and sternum. The postural changes are secondary to the chest deformity.
3. Operative correction of the chest deformity is described.
4. Post-operative physiotherapy is essential to correct the postural deformities. Patients must remain under orthopaedic supervision to maintain the correction obtained.
5. A series of twelve children treated by operation is reported, with excellent results in all.

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