The Operative Correction of Pectus Carcinatum (Pigeon Breast) *

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STERNAL DEPRESSION, such as seen in the deformity known as Pectus Excavatum, or Funnel Chest, is by all odds the commonest thoracic deformity to be treated operatively. There is now fairly widespread recognition of the fact that operation in patients with this deformity corrects the deformity from a cosmetic and orthopedic point of view, prevents progression of the deformity, and the development of cardiovascular symptoms, relieves cardiovascular symptoms when they are present, and, in many small children, results in obvious increase in vigor and growth. It is thought that the symptomatic effects of the deformity are due to compression and displacement of the heart, and perhaps to some compression and distortion of the lungs. The occasional occurrence of dysphagia is presumably due to pressure transmitted to the esophagus through the displaced heart.

The protrusion deformities of the sternum are substantially less common and their operative treatment on a less firm basis. While we have operated on, now, more than a hundred patients with pectus excavatum, we have treated only three patients with protrusion deformities of the sternum. The few papers on the subject of treatment of protrusion deformities of the sternum have been those of Chin,[2] Howard,[4] Lester,[5, 6, 7] and Brodkin.[1] There are two types of protrusion deformities, quite different in their appearance, probably quite different in etiology, and requiring different modes of operative correction.

The first, producing a pouter pigeon breast (Fig. 1), is undoubtedly entirely congenital. It is marked by a prominent forward tilting of the manubrium, with what amounts to a manubrio-gladiolar prominence. Distal to this the gladiolus inclines posteriorly, and actually forms a type of pectus excavatum deformity. Such a deformity we reported in 1952,[9] in a college boy who had repeated attacks of palpitation, followed by apparent slowing of the heart beat and heavy breathing. These attacks had occurred about once a month in the year preceding admission. An associated deformity was a failure of fusion of the laminae of the second thoracic vertebra. The deformity was corrected by resection of portions of five costal cartilages on the two sides, and by performing a cuneiform osteotomy at the manubriosternal junction, permitting the corpus sterni to be elevated forward, followed by a second osteotomy in the reverse direction in the mid-sternum permitting the distal portion of the sternum to be flexed posteriorly, correcting the concavity. This left the abnormally prominent manubrium undisturbed, but gently rounded out its continuation into the corpus sterni, at the same time correcting the depression of the distal portion of the sternum (Fig. 1B, D). Symptoms were relieved by this operation. I am indebted to the report of Currarino and Silverman, in 1958,[3] for

* Presented before the Southern Surgical Association, Hot Springs, Virginia, December 8–10, 1959.

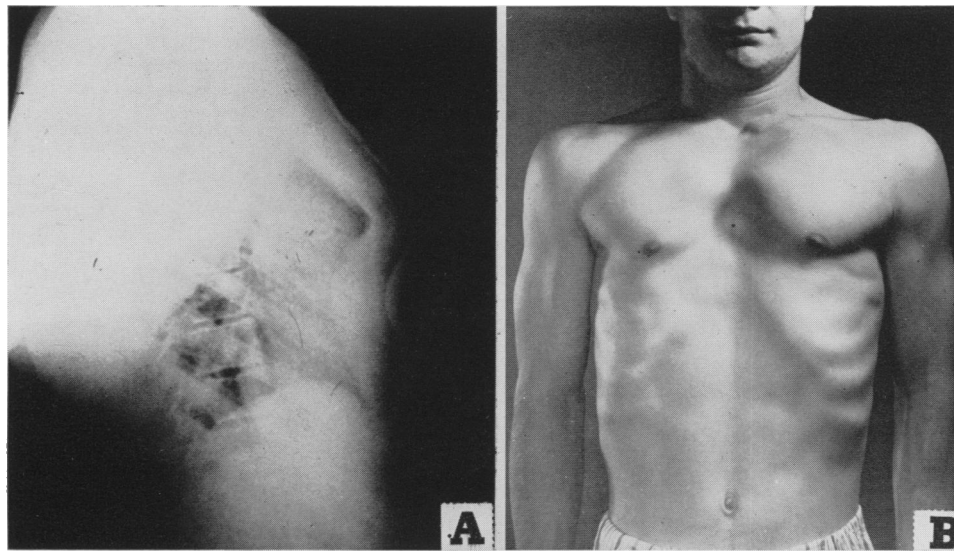

FIG. 1. B.R.—18-year-old college student, with attacks of palpitation followed by apparent slowing of the heartbeat, and heavy breathing. (From the Journal of Thoracic Surgery 23: 138–144, 1952) A and B a peculiar conformation of the chest—the manubrium slopes broadly forward to the manubrio-gladiolar junction, where there is a sharp posterior angulation. From this point the gladiolus inclines posteriorly to the lower third of the corpus sterni, where the slope reverses itself and the sternum tilts anteriorly. Both the photograph and the roentgenogram show the abnormal prominence of the manubrium associated with a depression of the corpus streni.

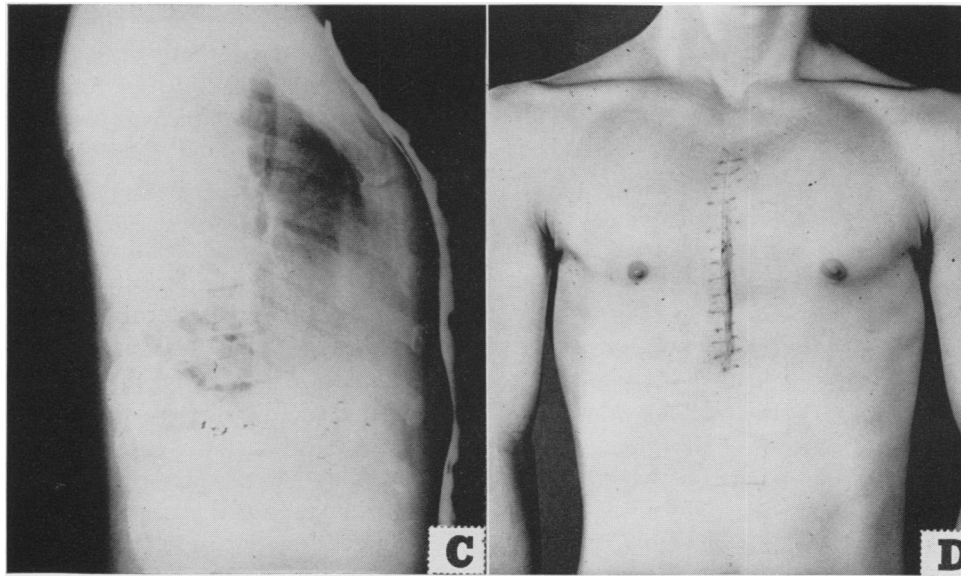

FIG. 1. C and D—After operation the chest is normally full and the roentgenogram demonstrates a smoothly continuous curve of the sternum. One of the corrective osteomoties shows plainly. Note that the lateral views of the sternum show no trace of the sternal segments. Currarino and Silverman point out that this type of deformity may be associated with premature fusion of the sternal segments.

pointing out that, in some instances at least, in the pouter pigeon breast deformity there is abnormal fusion of all the sternal segments. This certainly appeared to be true in this patient, and in another, whom we have seen, but upon whom we have not operated.

The deformity which perhaps is more commonly thought of as being that of pectus carinatum, or pigeon breast, similarly presents a considerable prominence of the sternum, but most of the prominence is of the corpus sterni, the manubrium being much less tilted forward, if at all. It has been our observation and the observation of several of those who have discussed this subject that a characteristic feature of these chests, is the lateral depression to either side of the sternum, accentuating the sternal prominence. It is my feeling that it is this lateral deformity toward which attention should be directed. This lateral depression may be quite deep enough to compress the heart and to reduce substantially the volume of the thorax. Correction of the two lateral depressions not only restores the volume of the thorax and relieves any possibility of compression of the heart or of the lungs, but restores the thoracic configuration to one that is essentially nearly normal.

The etiology of the various malformations which come under the head of protrusion deformities of the sternum is quite unknown. The pouter pigeon type of deformity seems usually to be quite fully developed at birth, and is probably entirely congenital. In the various protrusion deformities associated with lateral depressions of the costal cartilages, it seems difficult to avoid the feeling that the diaphragm plays an important part in the production of the deformity. Brodkin [1] believes that the various deformities of the sternum, both depression and protrusion, are the result of failure of the development of muscle in varying portions of the diaphragm, these portions then exerting a pull on the attached chest wall as a result of the unopposed action of muscle on the other side of the central tendon. Chin [2] is inclined to accept this explanation. It is certain that the lateral sulci in congenital sternal prominence are accentuated on deep inspiration. Whereas if there is any movement of the lower sternum on inspiration, it is outward. In the pouter pigeon deformity, the entire chest expands well. In funnel chest it is the sternum which moves inward on inspiration. The autopsy evidence which Brodkin, and Chin, use to support their theory is extremely slender. Chin has, however, reported biopsy of the central portion of the diaphragm in a number of instances of funnel chest, finding, at operation, an abnormal amount of fibrous tissue in the anterior portion of the diaphragm, the area incriminated in this theory, while the absence of muscular tissue in the more lateral portions of the diaphragm is held responsible for the production of the lateral depressions in pectus carinatum. Lester's 1953 report [5] was the first operative attack upon this problem to be published. He resected small segments

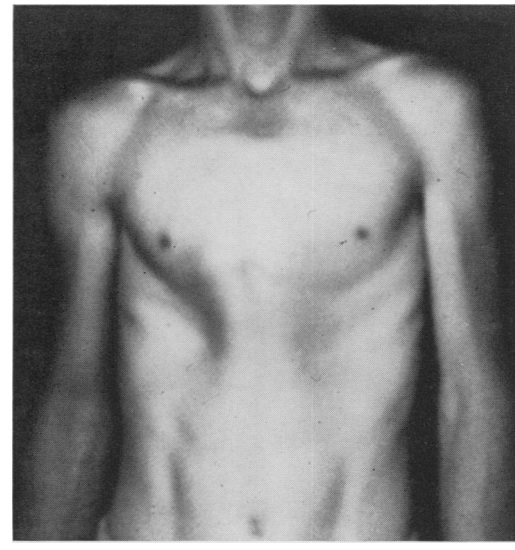

FIG. 2. W.P.—18 years old. Preoperative picture showing the unusual prominence of the sternum and the keel-like effect produced by the depressed areas in the costal cartilages on either side of the lower portion of the sternum.

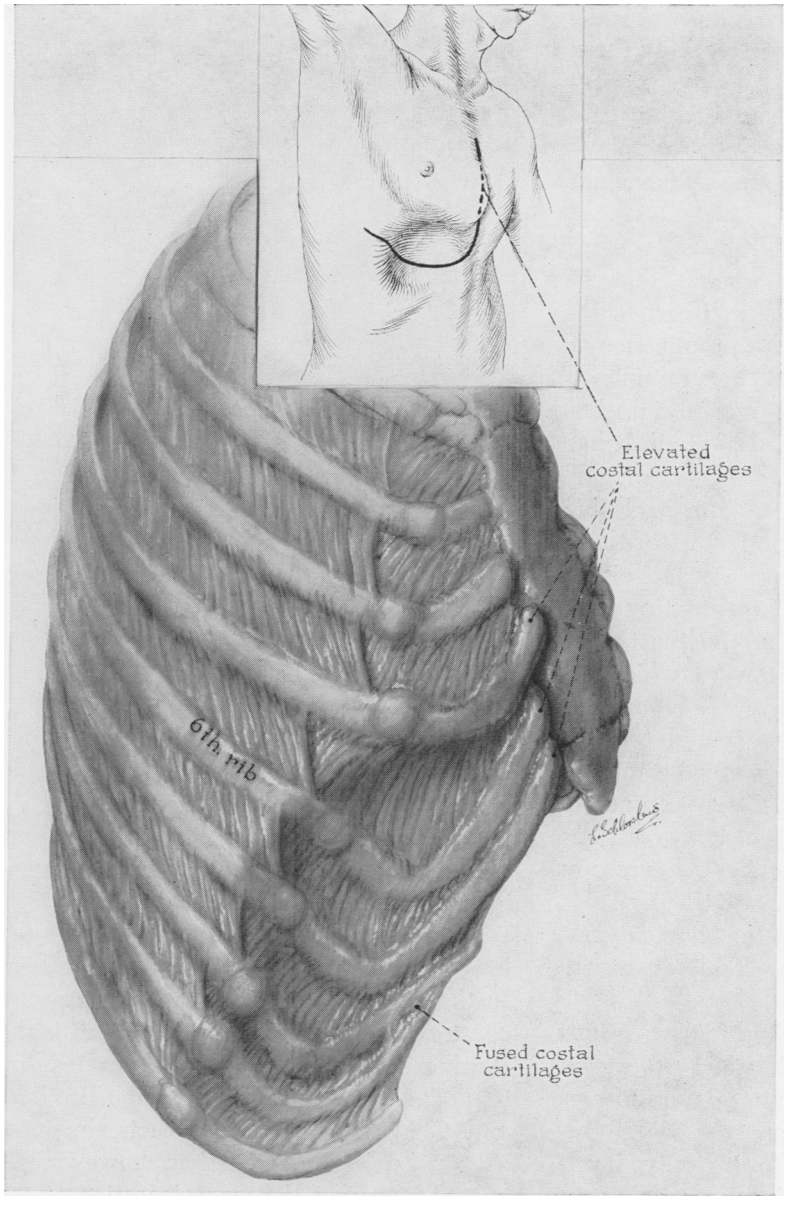

FIG. 3A. The operation for pigeon breast performed on W.P. This operation was performed in two stages for fear of the patient's not being able to tolerate a bilateral procedure. The operation in fact bothered him not at all, so that the subsequent operation on a four-year-old child was performed through a transverse incision and the entire operation completed in a single stage. The extent of the deformity shown, together with the incision employed.

of the costal cartilages subperichondrially, and then excised the entire corpus sterni subperiosteally. The sternum apparently regenerates in a depressed position after this procedure, a change which might not be thought advantageous. Nevertheless, a number of patients who were symptomatic before operation were relieved after operation. Chin,[2] in 1958, suggested that the anterior portion of the diaphragm, in patients with congenital sternal prominence, was attached not to the xyphoid and to the lower cartilages, but to the rectus abdominis. He pointed out that the Harrison's grooves are accentuated on inspiration, the lower sternum moves forward on inspiration, but that the rectus sheath dimples near the tip of the xyphoid on inspiration. All of this is presumably associated with over-development of the anterior-lateral portion of the

diaphragm. His operation, which is neat and original, consists of the resection of the medial portions of the sixth and seventh costal cartilages on both sides subperichondrially, detaching the xyphoid and reinserting it into a slot made for it in the sternum, at the level of the fourth costal cartilage. In small children this is said to pull the sternum back into proper position, with time.

Howard, of Melbourne, Australia, writing in 1958,[4] employed this maneuver of Chin's, but added to it the subperichondrial resection of all of the deformed costal car-

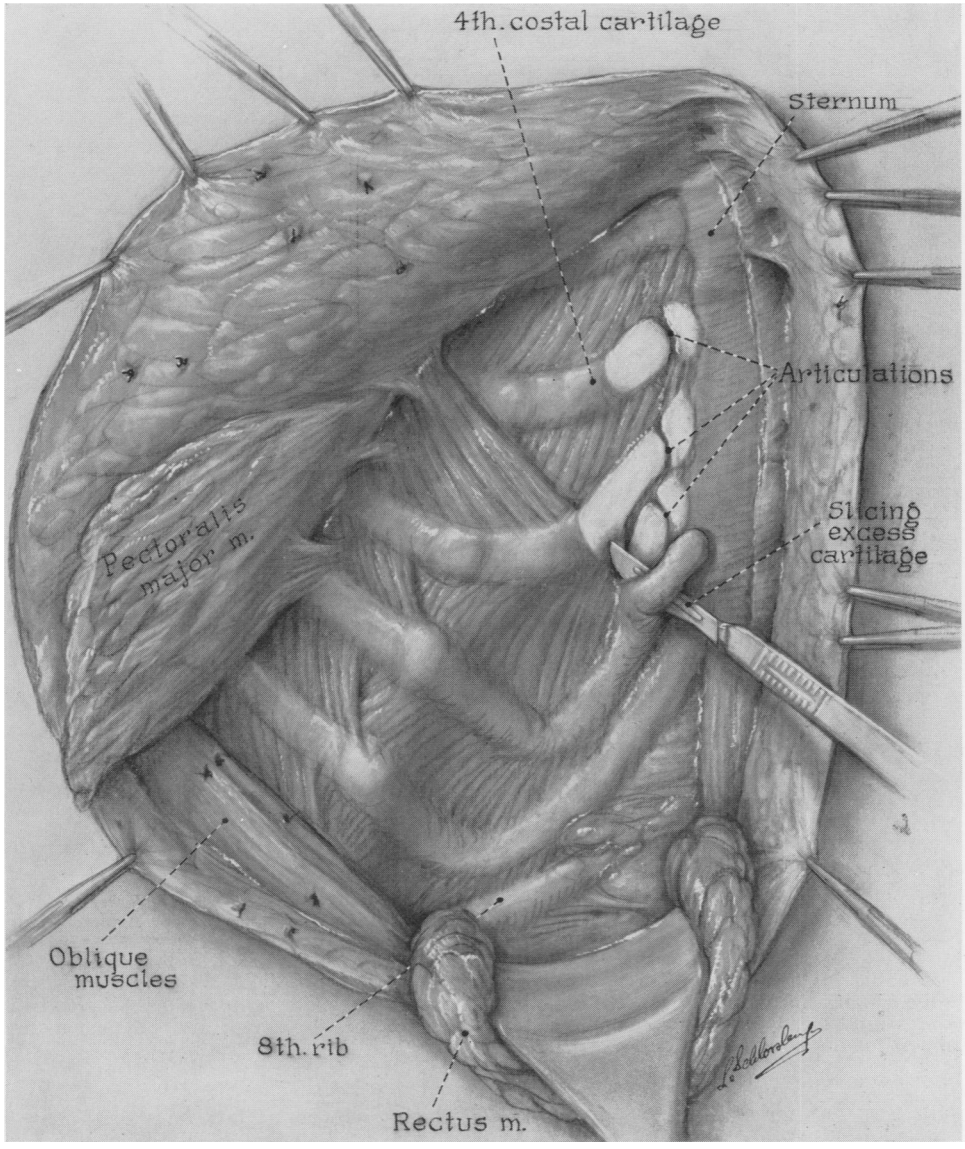

FIG. 3B. The pectoralis major has been reflected laterally and the rectus abdominis inferiorly. The prominence of the sternum had been exaggerated by knobby lesions of the costal cartilages. When these elevations were sliced away there were disclosed abnormal articulations in the fourth, fifth and sixth costal cartilages which appeared to be in the cartilages and not at the chondro-sternal junctions.

FIG. 3C. Involved portions of cartilages 5, 6, 7, 8 and 9 have been removed sub-periostally. In the instances of 6 and 7 the resection extends well back into the bony rib. Reefing mattress sutures are placed to straighten out the now redundant perichondrium and produce a taut support for the pleura in its newly expanded position.

tilages in the manner which we employed in our Case 2, in 1955. Brodkin has employed Chin's procedure.

In point of fact, there appear to be so many variations of protrusion deformities, even if we restrict ourselves to consideration of those in which the protrusion is predominantly of the sternum, that it is quite probable that a variety of operations will be required if these deformities are to be

satisfactorily corrected. I wish to present an operative approach which provides clear exposure of the deformity in that type of pectus carinatum associated with lateral depressions, and which allows for easy correction of the deformity. In all of these patients the depressed component of the deformity should be recognized, and it is this which should be corrected, by and large. It is hard to think that depressing the sternum can be good for these patients, and it might well be bad. The re-attachment of the xyphoid higher up on the sternum will hardly be [illegible] such as W.P., our Case 2.

Case Reports

W.P.—18 years old, was operated upon for the first stage on July 8, 1955, and for the second stage on March 23, 1956. This was a tall, gangling boy, with a pronounced cavity on both sides of the chest leading to an unusual prominence of the sternum, quite apart from the fact that his manubrium was sharply elevated (Fig. 2A, B). He claimed to be asymptomatic, and to wish this

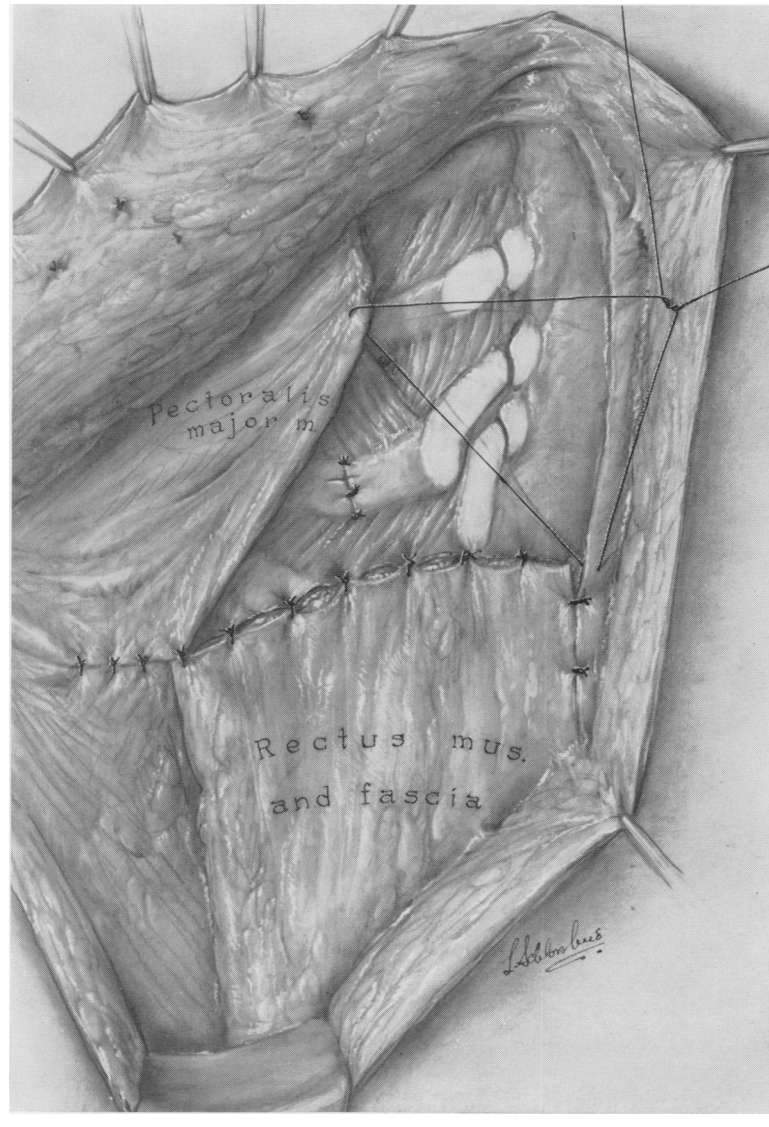

FIG. 3D. The pectoralis major and the rectus abdominis are sutured into place and the wound closed.

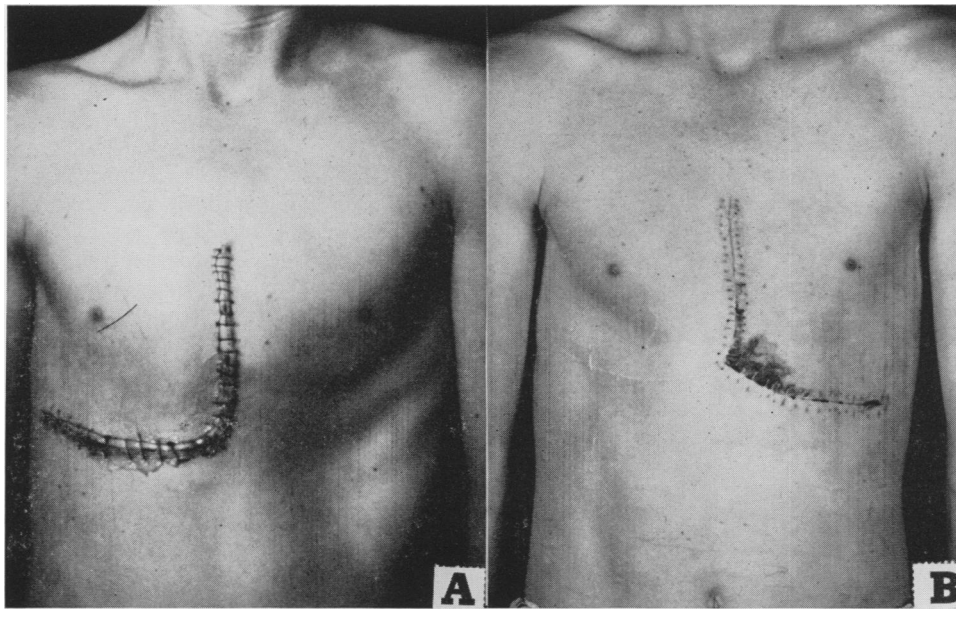

FIG. 4. W.P. The result after operation. A. After the first stage. B. After the second stage.

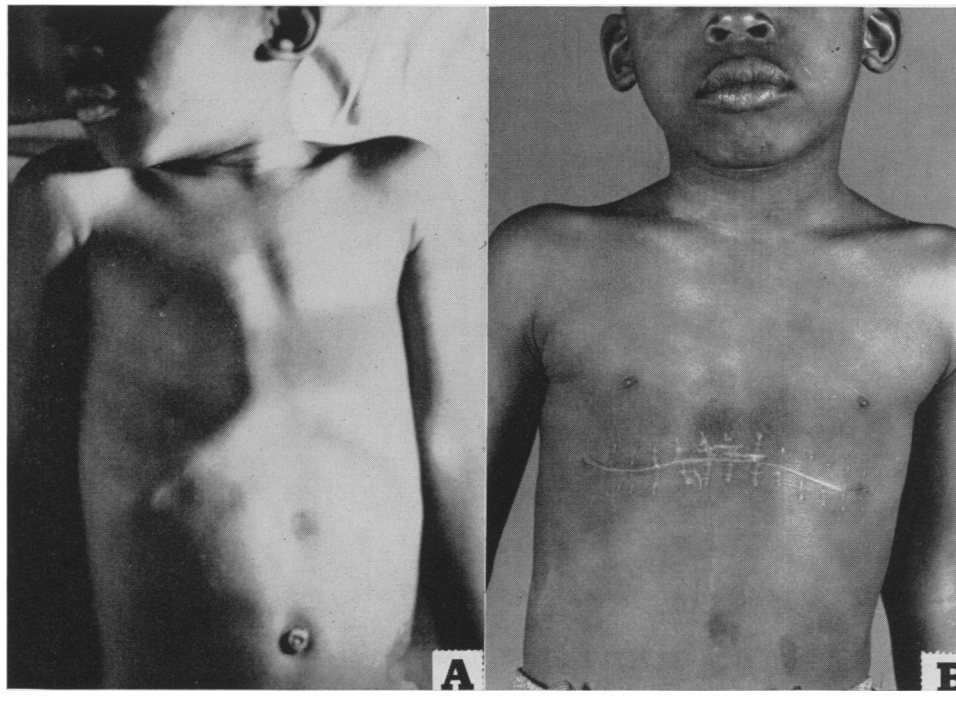

FIG. 5. M.J.—A four-year-old boy. The pigeon breast deformity present since birth, and thought to be progressing. A. The appearance before operation. The depression to each side of the sternum is quite striking. In the photograph the slope away from the sternum appears to be sharp, straight and on the right. It was in fact bilateral and concave. B. The appearance four months after operation. The transverse incision gave an excellent approach and an operation identical with that employed in patient W.P. was modified by the addition of an osteotomy for the distal portion of the sternum. The correction is complete.

operation only for cosmetic reasons. Following operation he stated that he was much freer in all of his activities, and felt better than he had for years. It is possible that he concealed symptoms from us in order to avoid any risk that we might refuse operation if he were not in perfect health.

At the first operation (Fig. 3, A, B, C, D) an incision was made running downward in the midline, and then across to the right axillary line, at the level of the lower end of the sternum, through the deepest point of the lateral concavity (Fig. 3A). The pectoralis major was reflected laterally, and the rectus abdominis elevated and reflected inferiorly, completely exposing the thoracic cage on the right. The fourth, fifth and sixth costal cartilages showed unusual prominences at their junction with the sternum, still further accentuating the medial prominence. These knobby prominences were cut away with the scalpel, disclosing unusual and odd joints in the costal cartilages (Fig. 3B). These gave further weight to the possibility that the deformity is, in part at least, the result of abnormal formation of the cartilages, or overgrowth of the cartilages. The five cartilages most deeply depressed were excised subperichondrially for the length of the depressed portions. It was striking that, as each cartilage was resected, the pleura lifted the perichondrium up into the wound, correcting the concavity. In order to maintain this correction, reefing sutures were taken in the perichondrium as shown in Figure 3C. This very nearly obliterated the concavity. The rectus and pectoralis were re-sutured (Fig. 3D) and the skin closed. The boy had a remarkably smooth convalescence, without any difficulty with respiration, and without much in the way of pain (Fig. 4A). On this basis we resolved that the next patient would be operated upon all in one stage.

Eight months later the second stage was performed on the left side. The same procedure was employed as before, with an equally gratifying result (Fig. 4B). On this side there were no anomalous chondral articulations.

On July 30, 1959, through the kindness of Dr. J. T. Hopkins, the opportunity to perform the one stage procedure came.

M.J. was a four year old boy. It was thought that his deformity had been present from birth, but had been growing worse (Fig. 5A). It was entirely similar to the deformity in patient No. 2. In this instance a transverse incision curved cephalad over the sternum, somewhat in the manner of a bow, gave access for a bilateral operation. In this patient portions of four cartilages on both sides were excised subperichondrially, and the same reefing sutures taken. In addition, in this youngster, the very distal portion of the sternum was found to be angulated posteriorly. A transverse cuneiform osteotomy across the distal sternum allowed correction of this angulation of the distal portion of the sternum. This child tolerated the bilateral operation very well, and, after a brief hospital stay, was discharged with a very satisfactory reconstruction of his chest wall (Fig. 5B).

Summary

Midline protrusion deformities of the sternum—pigeon breast—have two principal varieties. The first producing a pouter pigeon-like deformity consists of an undue prominence of the manubrium and a depression of the distal sternum, and may be associated with premature fusion of the sternal segments. It can be corrected by resection of the involved cartilages and appropriate sternal osteotomies. The second is associated with the protrusion of the entire sternum, frequently most marked some distance along the sternum from the manubrium, and is characterized by deep depressions of the costal cartilages to either side. It can be very satisfactorily treated by subperichondrial resection of all of the involved costal cartilages and shortening their course with reefing sutures in the perichondrium.

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Discussion

Dr. Paul W. Sanger: Dr. Ravitch is probably the most articulate man I know in speaking English. I have also heard him talk in Russian, French, and German, and he is equally clever in these languages. When he shows a movie, I am really intimidated at the thought of discussing his paper; however, I am delighted to see he is as conversant in discussing pectus carinatum (pigeon breast) as he has been in his writings on pectus excavatum (funnel chest).

When one stops to reflect, he must accept the fact that the chest has a very real function in aiding breathing. Actually it works in a bellows-like fashion. Its function is to move air in and out; therefore when a chest is in a frozen position, as it is in the deformity of keel chest, the individual is penalized, for he is not getting his fair share of oxygen into his lungs to be exposed to the gaseous exchanging elements. I think, however, the most distressing thing is that the patient is unable to move the air out, which eventually results in chronic destructive emphysema.

It is legendary that the demise of a person with kyphosis eventually will be caused by right sided heart failure or cor pulmonale. What Dr. Ravitch is speaking about tonight is in reality no more than an anterior form of kyphosis.

This is similar to the story of the Negro dean of a medical school who was giving his graduating class a last bit of advice. He said, "Now, gentlemen, you are going out to compete with white doctors, and one thing you must always remember is that when you establish a diagnosis, stick to it." Soon afterward, a young graduate had a very complicated case. The family was sitting around anxiously and expectantly when the young doctor came out and said very knowingly, "I am sorry to tell you that your son has the locked bowel." The mother then promptly replied, "He couldn't have, doctor; he has diarrhea." The doctor then quickly retorted, "I know, but it is locked in the open position."

Here as in the anterior kyphotic chest or pectus carinatum, we might say the chest is locked in an open position. We are convinced that this chest deformity develops from an overgrowth of the costal cartilages and until a free moving chest is created, this individual is doomed eventually to chronic emphysema, which if permitted to continue can result in right sided heart failure. It is agreed that the severe form of this condition should be operated upon, but the question is, "What is the severe form?" We believe that as soon as the patient has respiratory symptoms, his paradoxical breathing should be corrected.

We have operated upon 153 cases of anterior chest deformities, of which 139 were the funnel chest type. One of ten of the entire group was a pectus carinatum. We have adopted the philosophy that these cases should be operated on at a young age, as the deformity increases with growth, and it is very important to establish free motion of the chest for the youngster to breathe and develop properly.

Dr. Rollin A. Daniel, Jr.: I have been somewhat interested in the funnel chest deformities of children which, as you know, vary tremendously insofar as the asymmetry of various deformities are concerned and have been interested in Dr. Ravitch's belief that these deformities are of considerable functional as well as cosmetic importance.

I think, as Dr. Sanger has pointed out, that the keel breast type of deformity does lead to fixation of the chest wall and deformities of the spine as well as of the anterior chest wall and of fixation of the chest wall with consequent interference with ventilation of the lungs.

I have had but small experience with the keel breast deformity insofar as surgical therapy is concerned; we've operated on only three patients and have done essentially the same procedure as was shown by Dr. Ravitch, I think about the only difference being that we have divided the muscles and the perichondrium laterally and brought them external to the thoracic cage suturing these muscles and fascia and perichondrium under some tension maintaining the sternum in at least a nearly normal position.

This is essentially the same procedure that we've carried out for the funnel chest type of deformities.

Dr. Mark M. Ravitch: (closing) I appreciate the discussions of both Dr. Sanger and Dr. Daniel, both of whom have contributed so much to the surgery of this field and would entirely agree with them about the importance of performing these operations and performing them earlier.

With respect to linguistic prowess, I couldn't hope to match Paul in his ability to do Gullah dialect.