Evolving Management of Pectus Excavatum Based on a Single Institutional Experience of 664 Patients

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Most pediatricians and family physicians believe that children with pectus excavatum require surgery only for cosmetic indications and then only in teenagers. We believe pectus excavatum should be repaired in childhood (1) to relieve structural compression of the chest and allow normal growth of the thorax; (2) to prevent pulmonary and cardiac dysfunction in teenagers and adults; and (3) to obviate the cosmetic impact that may cause a child to avoid sports and gymnastics. Preoperative CT scans now help select those children who need repair to prevent progressive deformities. Pulmonary function studies during vigorous exercise can document respiratory dysfunction in teenagers. These features are reversible if repair is completed before the pubertal growth spurt. The ideal age for repair is 4 to 6 years, which permits enough emotional maturity for a positive hospital experience and avoids later psychological effects. Repair at an earlier age has no operative advantages. Our operative technique consists of the removal of three to four overgrown costal cartilages, repositioning of the sternum with a transverse osteotomy, and internal support using the child's lowest normal ribs, avoiding any prosthetic support. To prevent recurrence in teenagers, we add a temporary bar beneath the sternum to prevent depression of the sternum from the weight of the chest-wall muscle mass. Six hundred sixty-four patients have been followed for 1 to 40 years; 95% have excellent long-term results and only 5% have mild to moderate recurrences. Our current techniques of patient selection and surgery will be presented.

PECTUS EXCAVATUM (FUNNEL CHEST) is the most common chest-wall deformity seen by pediatricians and family physicians.[1] Most primary physicians believe that the repair of pectus excavatum is indicated only for cosmetic reasons and then only in teenagers. We believe this concept is wrong and recommend repair of significant pectus excavatum in childhood (1) to relieve structural compression of the chest and allow normal growth of the thorax; (2) to prevent pulmonary and cardiac dysfunction in teenagers and adults; and (3) to obviate the cosmetic impact that may discourage a growing child's participation in sports and gymnastics.[2] Our indications for repair and our surgical technique have evolved over 40 years and more than 650 cases. This experience from one institution forms the basis for our report.

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Clinical Review

During a 40-year period, 664 children, teenagers, and young adults had surgical correction of their pectus excavatum defects. The medical records and office charts of these patients were reviewed to document age at diagnosis and operation, radiographic findings, type of surgical repair, use of prothesis, and immediate as well as long-term results.

A total of 664 patients were evaluated and surgically treated for pectus excavatum. Pectus excavatum was seen more frequently in boys (498 patients) than in girls (166 patients), yielding a ratio of 3:1.

As seen in Table 1 the most common elective age for repair of the defect was just before or as the child entered elementary school (4 to 6 years). When the repair was performed later, during the teenage or adult years, this delay was due to late referral or worsening of a mild defect during pubertal growth. A few patients with the Marfan Syndrome were referred late for correction prior to open heart surgery.

The postoperative course of these 664 patients was remarkably uncomplicated, with a mean hospital stay of seven days and, in our most recent series, only five days.

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TABLE 1. *Age at Operation for 664 Patients with Pectus Excavatum*

Age at Operation	Series 1 (1949–70)	Series 2 (1971–80)	Series 3 (1980–85)	Series 4 (1985–88)	Total
2	38	0	0	0	38 (6%)
3–5	89	60	60	51	260 (39%)
6–14	38	55	71†	65†	229 (34%)
15+	18	26*	45*	48*	137 (21%)
Total	183	141	176	164	664 (100%)

* Adkins Strut.

† Ages 11–14 had struts.

There were no operative deaths from the repair.(In 1955, however, one infant died three weeks after pectus repair from continuing heart failure from complex congenital anomalies.) The morbidity rate was less than 5% and included superficial wound infection or separation, pneumothorax, subcutaneous fluid collection, shift in substernal strut, and pneumonitis. Since 1971, with the uniform use of the electrocautery, none of the 481 patients undergoing surgical correction has required a blood transfusion. Patients were examined at two weeks after surgery to monitor healing, and at 7 to 8 weeks to allow them to resume full physical activity. These series vary chronologically, but the patients were followed every 2 years after the first postoperative year until they had full growth of the chest wall.

The follow-up results in 460 of the patients are presented in four chronologic series ranging from 1 to 40 years (Table 2). Early in the series, during evolution of the surgical technique, poor results and reoperation occurred in 10% of the patients. The more recent series reveal only three poor results in 352 patients, of whom two patients were 11 and 12 years of age and for whom a temporary substernal strut was not used for postoperative sternal support. Therefore, with our current technique, operative results for pectus excavatum are quite satisfactory with greater than 95% of the patients having good to excellent results.

Discussion

The preoperative evaluation of the pectus excavatum deformity has evolved along with the surgical procedure. In the 1950 to 1970 era the evaluation was subjective and focused on measurement of anatomic severity. Diagnostic techniques included the use of barium strips on the chest defect with lateral chest roentgenograms, measurement of water volumes to fill the defect, obstetrical pelvimetry calipers to map the chest configuration, and Moire photography to define topographical changes of the chest wall.[1,3,4] Recently we[5] and others[6] have demonstrated the value of limited computerized tomography of the chest in the pre- and postoperative evaluation of pectus excavatum (Fig. 1). If the ratio of the transverse diameter to the narrowest anteroposterior diameter of the chest was >3.25, the children were found to have moderate to severe defects and required operative repair. In addition, all surgically treated patients had significant shifting of the heart and middle mediastinum into the left chest cavity. The most severe deformities had an additional rotation or twisting of the sternum, always to the right side (Fig. 1).

In the 1970 to 1985 era, physiologic methods of preoperative evaluation of patients with pectus excavatum have been attempted. These often require maximal exertion, invasive monitoring, radioisotopes, or significant patient psychomotor skills.[7–9] Because of the sophistication of these tests, most of these studies have been limited to teenagers and adults. However, recent exercise tests by Cahill[10] and Peterson[11] have clearly documented significant decreases in pulmonary and cardiac function during maximal exercise such as bicycle riding in children with severe deformities. With future technologic advances in measuring cardiopulmonary performance, such measurements in young children may allow further selection of children prior to their pubertal growth. In teenagers

TABLE 2. *Follow-up Results in 460 Patients (1988)*

Result	Series 1 18–39 Years	Series 2 9–17 Years	Series 3 4–8 Years	Series 4 1–3 Years	Total
Excellent	65	53	100	122	340 (74%)
Good	24	11	35	26	96 (20%)
Poor	9	0	2*	1	12 (3%)
Reoperation	10	2	0	0	12 (3%)
Total	108	66	137	149	460 (100%)

* May require reoperation—No bars.

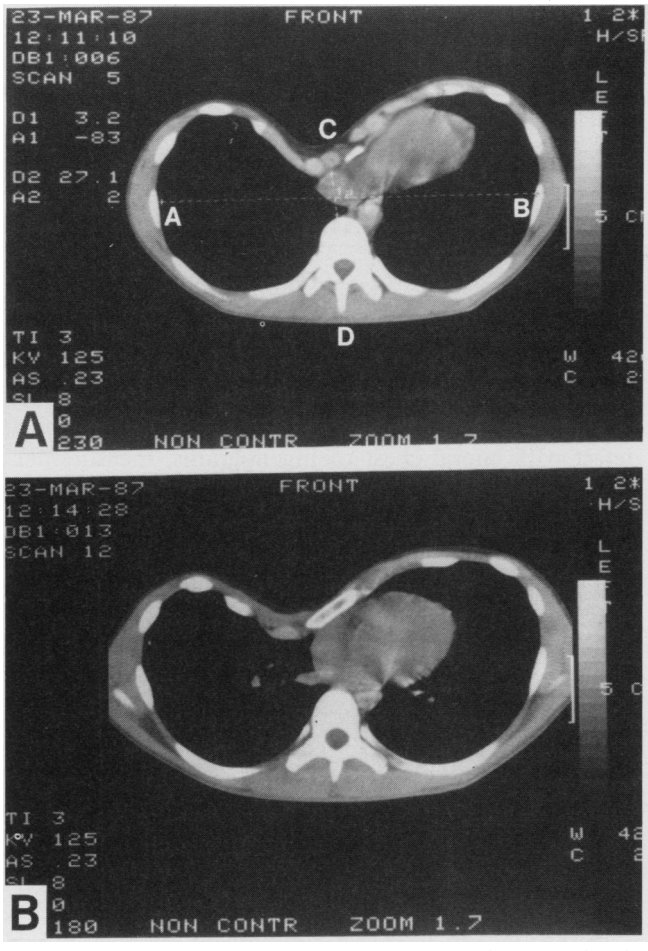

FIG. 1. Chest CT scan of pectus excavatum. (A) CT image at xiphoid with A-B (transverse diameter) and C-D (anterior–posterior diameter) indicated for calculation of pectus index A-B/C-D. (B) CT image demonstrating angulation of sternum to the right in the severe defect.

the tests are simpler and strongly positive; indeed, many teenagers complain of shortness of breath and fatigue with strenuous exercise.

Hecker[12] in Munich and Welch[13] in Boston have reported two series of patients using similar surgical methods without the use of substernal stabilization in children and both report good to excellent results in more than 90% of their patients. One of the largest series, by Willital,[14] reports the use of substernal struts in over 1000 patients with a satisfactory to excellent result occurring in 92% of the patients. In our series, the use of struts was selective and temporary. In our earlier series, patients older than 12 years of age had a substernal strut placed for stabilization of the sternum, as well as the basic tripod fixation. This was based on findings of our earlier series that the teenager's chest wall musculature was heavier and required further support of the lower portion of the sternum to prevent recurrence. Early in the third series, two children, ages 11 and 12 years, had recurrences with only tripod support. Since then, children 11 years of age and older have been treated with temporary substernal stainless steel bar stabilization, and there have been no further recurrences. Of the large series in this country, only Welch and Shamberger currently use the modified Ravitch procedure[1] that we champion.

The two major advances in postoperative management have been the use of substernal and subcutaneous drainage, which has decreased subcutaneous fluid collections from 40% to less than 5% and subcuticular skin closure, which gives a much nicer cosmetic scar than formerly used skin sutures.

Thus, evolution in the management of pectus excavatum over the past 40 years has resulted in a safe, standard procedure that yields satisfactory results in more than 90% of the patients.

Essential Features of the Surgical Repair of Pectus Excavatum

The operative technique that has evolved over the past four decades is a gradual modification of a technique developed by Dr. Mark Ravitch that consists of the following basic procedures.

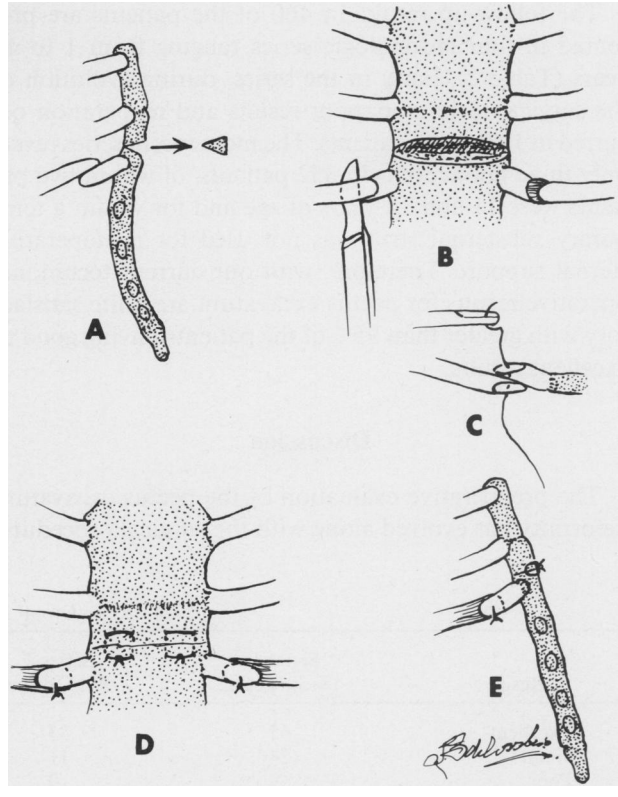

FIG. 2. Tripod fixation of the sternum. (A) Oblique chondrotomy of the lowest normal rib. (B and C) Overlap and fixation of the tripod. (D) After cuneiform osteotomy, sternal elevation and fixation through the adjacent periosteum.

(1) Transverse skin incision below the nipples (submammary in girls) and centered, if possible, over the deepest part of the concavity.

(2) Elevation of skin flaps at the level of the pectoral fascia using electrocoagulation to minimize blood loss.

(3) Reflection of the pectoralis muscles by incising the fascia in the midline and sweeping the muscle flaps laterally with an inferior transverse relaxing incision along the seventh rib.

(4) Subperichondral removal of all abnormal costal cartilages (usually ribs 4 through 7 bilaterally), carefully preserving the perichondrium.

(5) Subxiphoid incision and finger mobilization of the substernal space, avoiding entry into the pleural spaces.

(6) Transection of the intercostal muscle bundles from the sternum to permit full mobilization of the sternum.

(7) Oblique transection from medial to lateral of the costal cartilage of the lowest (third or fourth rib) normal rib (Fig. 2).

(8) Transverse anterior cuneiform (wedge) sternal osteotomy just above the lowest normal rib (Fig. 2).

(9) Elevation and fixation of the sternum with mattress nonabsorbable sutures through the periosteum at the site of the osteotomy. With sternal elevation, the medial part of the transected cartilage lies on and is supported by its lateral partner and is then fixed with a single suture (Fig. 2).

(10) The sternal fixation and bilateral sternal support constitute the internal dynamic tripod suspension and obviates the need for additional hardware support.

(11) For children older than 10 years of age, teenagers, and adults, placement of a temporary stainless steel strut beneath the sternum is required and the strut is anchored bilaterally to the fifth or sixth ribs.

(12) Placement of a soft plastic chest catheter in the substernal space.

(13) Reapproximation of pectoralis muscle fascia in the midline.

(14) Placement of small silastic drainage catheters beneath the skin flaps.

(15) Skin closure with subcuticular suture and plastic strips.

In the recent past (from 1980 to 1988) we have identified a special group of young adults whose indication for repair is correction to permit a median sternotomy for open heart surgery. These are the Marfan Syndrome patients who need aortic root replacement and/or aortic and mitral valve repair.[15] Their sternums are so severely depressed and rotated that the heart is displaced into the axilla (Fig. 3). A median sternotomy is technically difficult, if not impossible, and cannulation of the displaced heart for bypass is hazardous. In addition, we believe postoperative pulmonary function is greatly improved by earlier correction of the pectus deformity. Five patients with

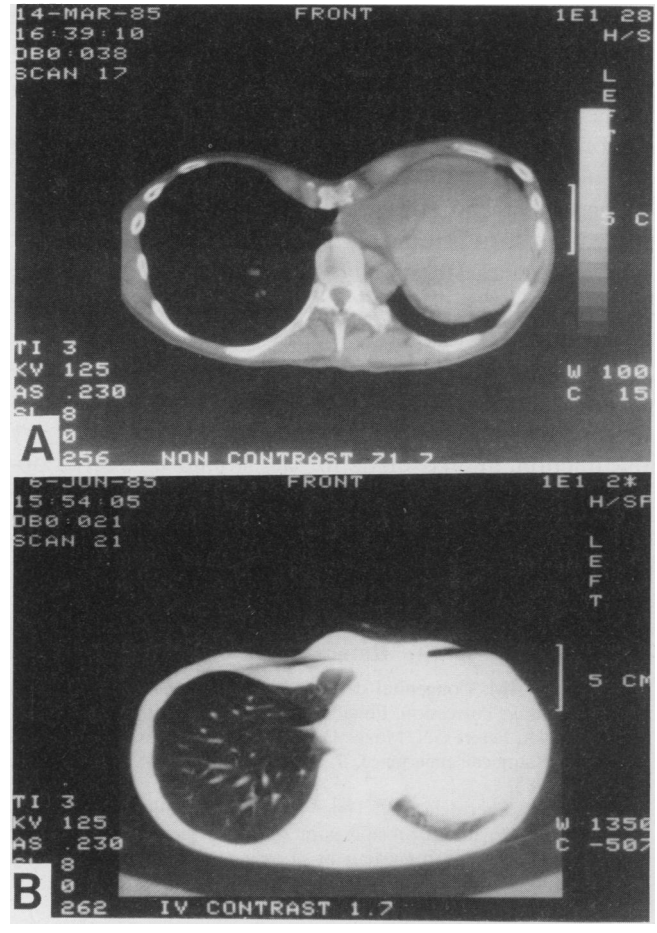

FIG. 3. Pre- and postoperative chest CT of pectus excavatum repair. At the same level of the defect, (A) preoperative and (B) postoperative views.

Marfan's Syndrome have been managed successfully with staged pectus and open heart surgery.

Although there are still no absolute criteria for operative intervention, our preference is to observe these children through early childhood and evaluate the progression of chest-wall growth and development. We carefully document any interference with the lower chest-wall rib growth, depth of defect, posture and cross sectional dimensions of the chest by serial caliper measurements. By five to six years of age it is usually clear which children have a moderate to severe defect and would benefit from surgical repair. At this age, the children are emotionally mature enough and are more likely to derive a positive experience from their hospital stay. This age is also early enough that most children have not suffered psychologically from peers due to cosmetic concerns. Finally, these children are old enough to protect themselves from "roughhouse" activities during the 6 to 8 week postoperative healing period that is necessary for rib cartilage regeneration and a solid chest wall. Presently the only precaution used postoperatively is the avoidance of physical contact activity for 6

to 8 weeks in children and for 8 to 10 weeks in teenagers. This time period is required for new cartilaginous rib growth from the perichondrium and secure attachment of the cartilage to the normally positioned sternum.

For the selected older children and teenagers requiring the placement of a substernal strut, the postoperative management and release to full physical activity is the same. The strut is removed on an outpatient basis under light general anesthesia by opening the lateral few inches of the previous incision. This is done 6 to 9 months after the primary repair.

On the basis of our experience and that of other surgeons, we believe that children who suffer from a pectus defect with its structural compression, cardiopulmonary dysfunction, and cosmetic impact should be offered operative repair. With current diagnostic and operative techniques, the pectus excavatum deformity can be safely repaired with full expectation of excellent long-term results.

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Discussion

DR. MARK M. RAVITCH (Pittsburgh, Pennsylvania): This is a good experience. Dr. Haller quite rightly emphasized the point that this deformity imposes a definite physiologic abnormality on these patients, and that the degree of abnormality that is determinable by our clinical laboratory examination is not necessarily proportional to the symptoms the patient has.

It is uncommon for the younger patient to have any symptoms at all, although we have seen dysphagia in newborns and have operated on them with success.

Dr. Haller is quite right. We operate any time we recognize the significant deformity, if the mother doesn't faint away when operation is proposed.

In about 1948 at the University Surgeons I presented the history of a man with his second bout of cardiac failure and atrial fibrillation with a severe pectus deformity. We showed a few things in him by the physiologic studies that we could do then, notably a right ventricular pressure curve not unlike the one in congestive failure with ascites.

At that time Herb Meyer, a very astute clinician, said after we pointed out that by and large we never found anything abnormal by cardiovascular catheterization or respiratory function test, that "When you develop more sophisticated and more accurate examinations and examine the right things, you will," and of course, that has now happened in examinations done all over the world.

We have minor points of difference with Dr. Haller in addition to the age of operation. I think it is easier to do the operation on small children. I think the fact that they don't anticipate the operation is great. I think most all of you would like not to anticipate operations. The operation does not seem to cause any particular trouble because of age of the small patients.

He points out in the manuscript that for years he hadn't used blood at all, and we first discovered this in the little babies and finally as we got older and better for a while, until we reached the other side of the curve, we got to the point where we just didn't use blood on anybody. This can be quite a bloodless operation.

We still like the midline incision, which we think gives better access and is no less unsightly. You had better predict to your patients or their families that you have no control over the healing. There may be a keloid and they may be sorry they had the operation done because the scar is so unsightly.

We have experimented with a variety of methods of closure that haven't seemed to make any difference.

I am interested that Dr. Haller believes that the disturbances of respiratory function have been better documented than the disturbances in cardiac function. I believe myself that the cardiac problem is probably the more severe. You almost always have compression of the right ventricular outflow tract. That has been shown over and over again, and it shows beautifully in the CAT scans.

As for the studies that we have been doing prior to operation–x-ray and physical examination. We have got enough CAT scans, angiograms and so on to illustrate any number of articles or talks. They are very costly. A thousand dollars or so to add to my slide collection seems an unfair burden on Medicare, the patient, Blue Shield, or whomever, and I don't think that it adds to the result.

About the only time in recent years that we have been using physiologic testing has been in adults who we thought claimed an exaggerated physiologic deficit and on whom we are unwilling to operate unless we could prove that there was such a deficit and because there is likely to be something whacky about somebody who waits until he is 25, 30, or 35 years old before coming to you for an operation like that. He is looking for success on the beaches, in the board room, in the bedroom, or wher-