6.3 Thoracic wall deformities

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6.3.1 Introduction

The open technique or Ravitch procedure for the repair of pectus excavatum involved radical resection of cartilages and "short ligaments" and was the procedure of choice until Pena in 1990 and Haller in 1996 [1, 2] drew attention to development of acquired asphyxiating chondrodystophy as a result of this extensive and early resection of the anterior chest wall structures. This was followed by the introduction of the minimally invasive bracing procedure by Nuss et al. in 1997 [3]. The technique has been compared to placing braces on the teeth. It requires no rib, cartilage, or sternal resection and consists of placing a curved steel bar under the sternum using thoracoscopy for guidance [3–5]. The procedure has since gained acceptance by the surgical community [6–10] because of good to excellent long-term results in over 95% of patients.

6.3.1.1 Indications for surgery

During the latter half of the twentieth century it was common practice for surgeons to operate on very young pectus excavatum patients based simply on their clinical evaluation [11, 12]. Since publications by Haller et al. and Martinez et al. drew attention to the risk of acquired asphyxiating chondrodystrophy [1, 2], most surgeons who still use the open technique wait until after puberty to perform the operations. Although the minimally invasive procedure can be performed at any age without risk of chondrodystrophy, the optimum age is just before puberty since the chest is still soft and malleable, the patients show quick recovery, the results are excellent and the risk of recurrence is very low. Patients who have their repair at a younger age and who have their bar removed before puberty are at a slight increased risk of recurrence.

The decision for surgical repair of a pectus excavatum not only relies on the history and physical exam but also strict objective criteria based on the results of the chest CT, pulmonary function tests and cardiac evaluation, including electrocardiogram and cardiac echocardiogram (Fig. 1). Anterior chest wall malformations can range from mild to severe and at our institution less than half the patients are severe enough to warrant surgical correction. Cardiac and/or pulmonary compression causes symptoms of dyspnea on exertion, chest pain with or without exertion and lack of endurance. Pul-

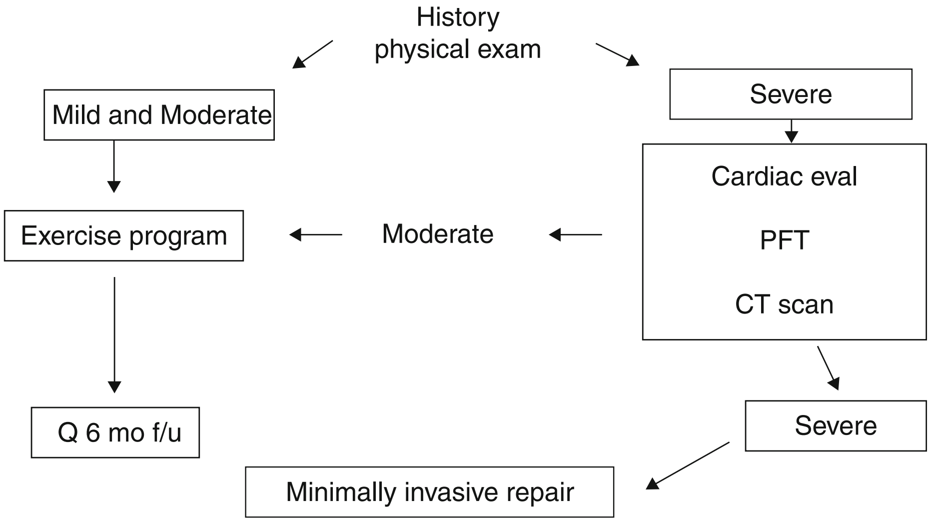

Fig. 1. Pathway for evaluation and treatment of pectus excavatum using the minimally invasive technique

monary function tests often show a restrictive or obstructive deficit. In addition to physical symptoms, a severe deformity may result in poor body image that impacts self-worth [13]. Patients undergoing rapid growth need to be re-evaluated at regular intervals since the deformities tend to progress as the patients grow. This can be especially noticeable during the pubertal growth spurt when a pectus excavatum can progress from mild to severe in as little as 6–12 months.

Classification and treatment algorithm

Initial evaluation of the patient requires a complete history and physical examination including photographs to document the pectus excavatum. Patients are then classified into mild, moderate, and severe categories. The patients with a mild or moderate deformity who are asymptomatic are started on an exercise and posture program and re-evaluated at 12-month intervals. The exercise program is initiated to improve cardio-pulmonary function, chest expansion and to strengthen the chest and back muscles in order to halt the progression of the deformity. Approximately 66% of the patients from our community are treated conservatively. The patients with a severe deformity and patients who have documented progression of their deformity undergo a workup to see whether they have objective evidence to support the need for surgical repair. The workup includes pulmonary function studies, a cardiology evaluation by a cardiologist, including an electrocardiogram and echocardiogram, and finally, a chest CT scan. For consistency, radiologists have suggested that the chest CT be performed during quiet respiration, not during maximum inspiration (Fig. 2). The need for surgical correction is determined if the patient has two or more of the following criteria: (1) a Haller index of greater than 3.25 demonstrated by CT scan; (2) cardiology evaluation demonstrating cardiac compression or displacement (Fig. 3) mitral valve prolapse, murmurs or conduction abnormalities; (3) pulmonary function studies showing restrictive and/or obstructive lung disease; (4) progression of the deformity with associated physical symptoms other than isolated concerns of body image; (5) recurrent pectus excavatum after a failed previous repair by a Ravitch procedure or a minimally invasive procedure.

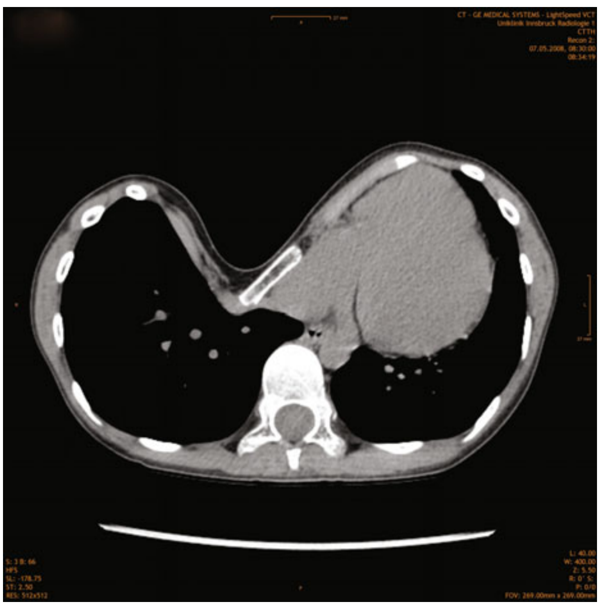

Fig. 2. CT scan showing severe pectus excavatum performed during quiet respiration

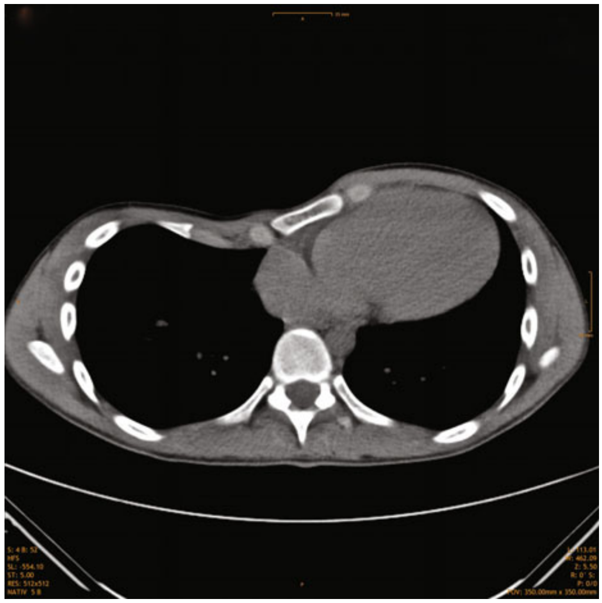

Fig. 3. CT scan demonstrating cardiac compression and displacement, as well as pulmonary compression

Preoperative assessment should also include asking about a history of metal allergy in the patient and immediate family. Nickel and Cobalt are components of the surgical steel used in the bar and are the cause of allergy in 2% of patients [14]. If a metal allergy is suspected, testing by a T.R.U.E. patch (Allerderm, Phoenix, AZ) can clarify this issue. In the presence of metal allergy, a non-allergic Titanium bar is used instead. Unlike the stainless steel bar, which is bent by the surgeon at the time of operation, titanium, must be bent in advance at the factory using CAD/CAM technology.

6.3.1.2 Exercise program

Most patients with pectus excavatum lead a very sedentary lifestyle and have a classic “pectus posture” which aggravates the deformity and may lead to progression of the deformity. Patients who are diagnosed as having a mild or moderate deformity are started on an exercise and posture program designed to improve cardio-pulmonary function and improve posture. Exercise can also increase chest expansion. Breathing and posture exercises are taught to the patients and they are instructed to do these on a daily basis. Patients are also encouraged to participate in aerobic activities such as team sports that involve swimming, running, etc.
Patients are re-evaluated at 6- to 12-month intervals to monitor compliance with the exercise program and to check progression of the deformity. It is possible to halt the progression in a mild deformity and slow the progression in a moderate deformity.

6.3.2 Surgical repair

Although the minimally invasive procedure can be performed at any age, the optimum age is just before puberty. The chest is still soft and malleable before puberty and the patients show quick recovery, a rapid return to normal activity, and have excellent results. The surgical correction of the pectus excavatum is accomplished by inserting a convex steel bar under the sternum with the convexity facing posteriorly. When the bar is in position, it is turned over 180° thereby correcting the deformity. The technique is made possible by the malleability and flexibility of the anterior chest wall and requires no cartilage incision or resection and no sternal osteotomy. After puberty the flexibility of the chest wall decreases making the repair more difficult and patients frequently require the placement of two bars. At our institution we have performed the procedure up to age 31 with equally good results [4]. Other authors have obtained excellent results in adult patients up to 50 years of age [15, 16].

6.3.3 Surgical technique

The preoperative checklist on the day before surgery includes reviewing all studies, checking for allergies including allergy to nickel, and measuring the chest to determine bar length. A measurement from the right to left mid-axillary line is taken and then 2 cm (±1 in.) is

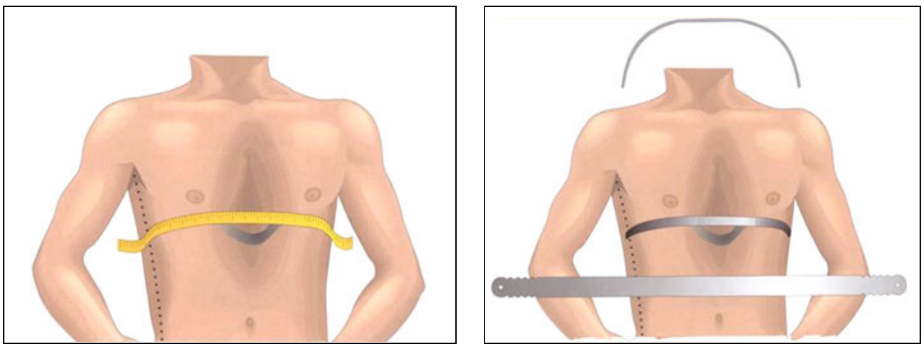

Fig. 4. Technique of measurement determining bar length. The bar needs to be 2 cm shorter than the measurement

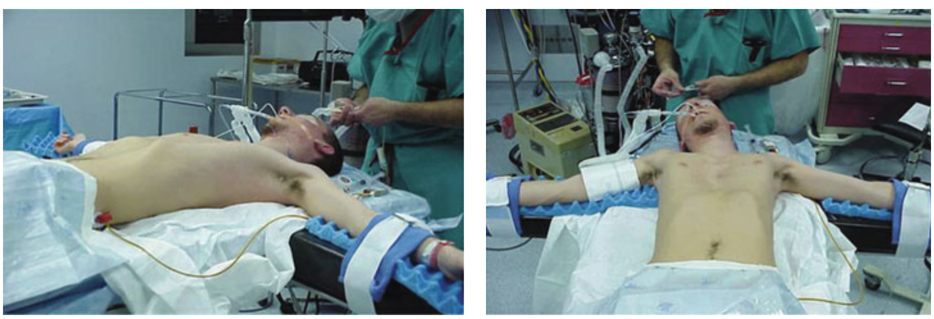

Fig. 5. Positioning of the patient to minimize brachial plexus injury

subtracted from this measurement. The bar takes a shorter course than the tape measure and consequently needs to be 2 cm (±1 in.) shorter than the measurement (Fig. 4).

General endotracheal anesthesia and epidural catheter insertion is undertaken by the anesthesiologist. The epidural catheter infusion is continued for 3–5 days with the average at our institution being 3 days. An indwelling bladder catheter is placed and this remains until postoperative day 1 at which time it is removed. Antibiotic coverage is provided and continued to discharge to minimize the risk of pneumonia with subsequent bar infection. Both arms are abducted at the shoulder and gel pads are used to prevent brachial plexus injury (Fig. 5). There should be slight flexion at the elbows. The patient is draped and the Lorenz™ pectus support bar is bent into a semi-circle, leaving the central 2 cm flat to support the sternum (Fig. 6). Bending the bar into an arch shape allows sustained load bearing of the bar. If the central flat section of the bar is too long, there will be undercorrection of the pectus excavatum.

Fig. 6. Demonstration of a properly bent pectus support bar. The middle bar is correctly bent. The upper bar is too flat, while the lower bar is too rounded

Marking the patient requires determining the deepest point of the pectus excavatum and marking this area with a surgical marking pen (Fig. 7). If the deepest point of the pectus is inferior to the sternum, then the inferior end of the sternum is marked instead. This point sets the horizontal plane for bar insertion. The intercostal spaces that are in the same horizontal plane as the deepest point of the pectus excavatum are marked with an "×". These thoracic entry and exit points on each side of the sternum should be medial to the top of the pectus (costochondral) ridge. Lines are drawn for the proposed incision sites on each lateral chest wall in the same horizontal plane (Fig. 7).

A thoracoscope is inserted through the right lower lateral chest wall approximately two interspaces inferior to the proposed right lateral thoracic wall skin incision (Fig. 8). A thorough inspection of the right hemithorax and mediastinum is performed checking that there is no contraindication for repair. Then pressure is applied to the intercostal spaces marked for bar insertion to ensure that the external markings line up well with the internal anatomy. After confirming by thoracoscopy that the internal and external anatomy match up well, bilateral thoracic skin incisions are made in the region of the mid-axillary line and a skin tunnel is raised anteriorly toward the intercostal space marked with an "×", medial to the top of the pectus ridge (Fig. 9). Also, a pocket is created for the distal end of the pectus bar and stabilizer. Under thoracoscopic

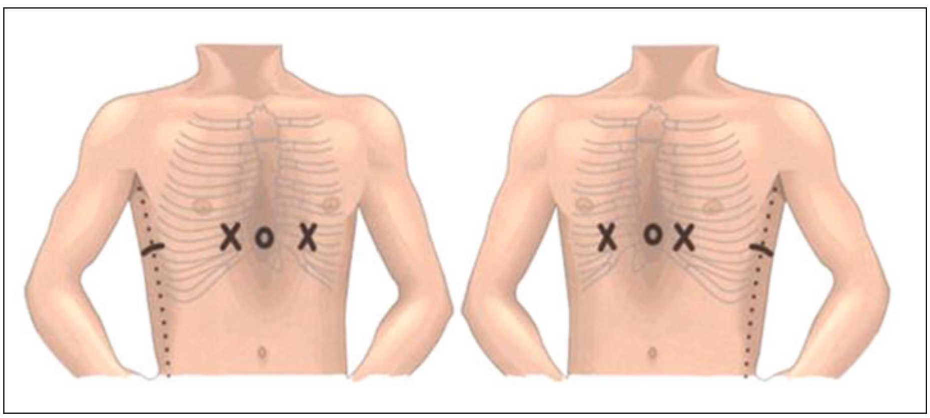

Fig. 7. Marking the bar insertion site. The skin incision, entry and exit sites and the deepest point of excavatum are all in the same horizontal plane

Fig. 8. Placement of thoracoscope for minimally invasive repair

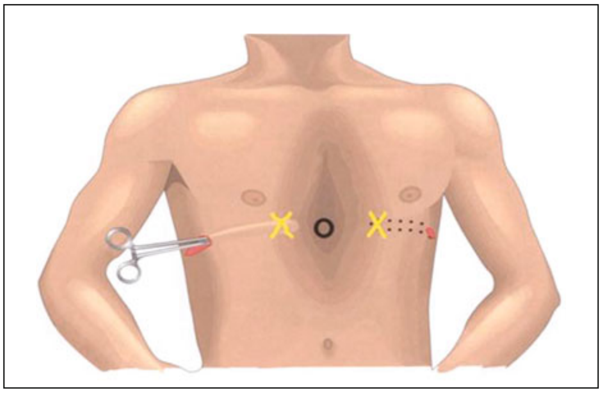

Fig. 9. Creation of the skin tunnel

control, the appropriate size Lorenz™ introducer (Biomet Microfixation, Jacksonville, FL) is inserted through the right intercostal space at the top of the pectus ridge at the previously marked "×" (Fig. 10). The EKG volume is turned up so that the heartbeat is clearly audible. The pericardium is gently dissected off the under surface of the sternum (Fig. 11). The introducer is *slowly* advanced across the mediastinum under thor-

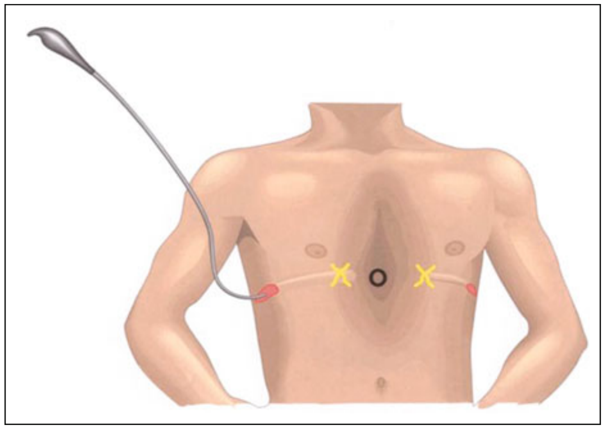

Fig. 10. Insertion of the introducer

acoscopic guidance with the point always facing anteriorly and in contact with the sternum. When the substernal tunnel has been completed, the tip of the introducer is pushed through the contralateral intercostal space at the previously marked "×", and advanced out of the skin incision (Fig. 12).

The introducer is then used to elevate the sternum. The surgeon lifts the introducer on the right side and the assistant lifts on the left side (Fig. 12). The lifting is repeated until the sternum has been elevated out of its depressed position and the pectus excavatum has been corrected. An umbilical tape is attached to the introducer, which is slowly extracted, pulling the umbilical tape through the substernal tunnel (Fig. 13). The previously prepared pectus bar is tied to the umbilical tape and guided through the substernal tunnel using the umbilical tape for traction and the thoracoscope for vision. The bar is inserted with the convexity facing posteriorly. When the bar is in position it is rotated 180° using the bar flipper (Fig. 14).

If the bar requires further bending, it is turned over and molded where required using the small Lorenz bar bender. If one bar is not enough, then a second bar is inserted

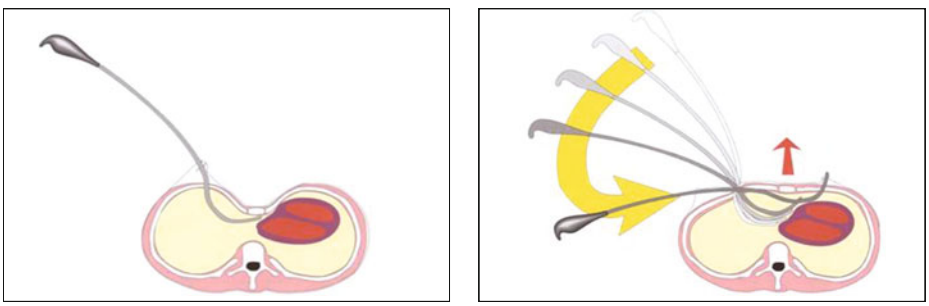

Fig. 11. Technique of dissection with the introducer

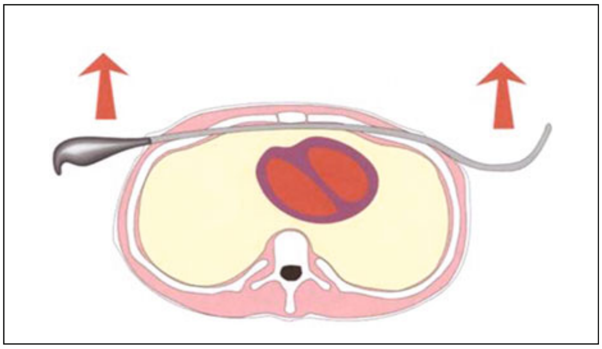

Fig. 12. Demonstration of placement of introducer and subsequent lifting on the sternum

one interspace above or below the first one. Two bars give better and more stable correction, especially in older patients. Slight overcorrection is necessary to prevent recurrence after the bar is removed. Some surgeons prefer approaching the mediastinal tunneling from the left rather than the right side. Other surgeons make a third incision over the xiphoid and then apply a towel clip to elevate the sternum and others do a finger dissection under the sternum before inserting the introducer [17].

6.3.3.1 Stabilization of the bar is absolutely essential for success

A stabilizer is inserted onto the left end of the bar and wired to the bar with No. 3 surgical steel wire. If the bar does not seem stable, a second bar rather than a second stabilizer is probably required. Heavy absorbable pericostal sutures of "0" or "1" PDS are placed around the bar and underlying rib using an "endo-close" laparoscopic needle under thoracoscopic vision on the right side (Fig. 15). Hebra and Gauderer advocated placing a suture adjacent to the sternum [18]. Once the bar is stabilized, the incisions are closed in layers and the pneumothorax is evacuated using the trocar and attached tubing (or a chest tube), with a "water-seal" system. A chest X-ray is obtained before the patient is taken out of the operating room, to check for a residual pneumothorax.

6.3.4 Postoperative management

In the recovery room the patient is kept well sedated with the goal of a smooth emergence from anesthesia.

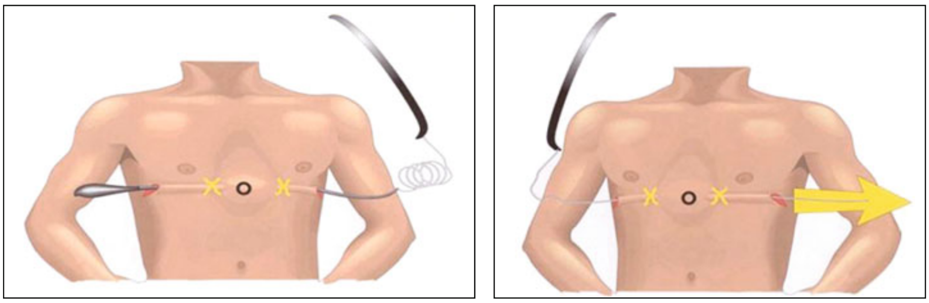

Fig. 13. Technique of using umbilical tape to guide the pectus support bar through the substernal tunnel

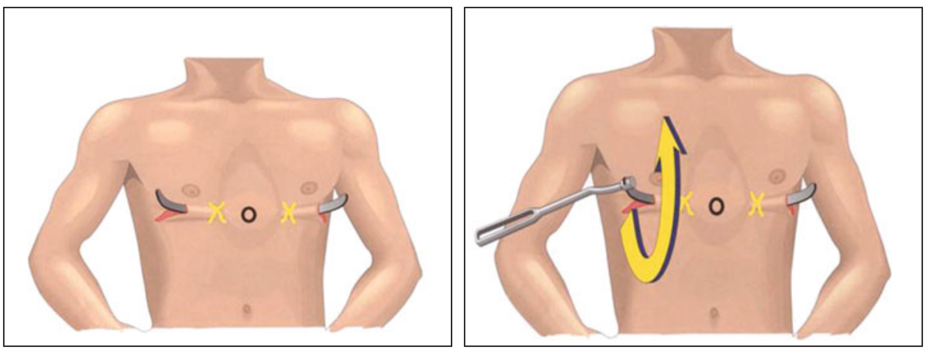

Fig. 14. Rotation of the pectus support bar using the bar flipper. Rotation may be clockwise or counter clockwise

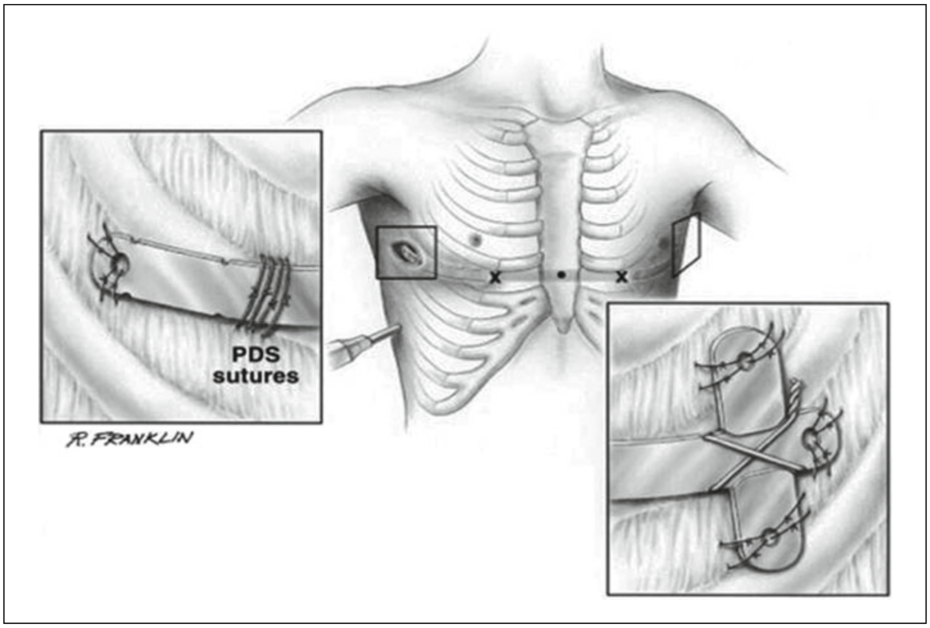

Fig. 15. Stabilization of the pectus support bar

The epidural catheter is left in for 3–4 days. The patient is discharged home on the fourth or fifth postoperative day. Patients may return to school after 2 weeks, but may not participate in sports for 6 weeks from the time of surgery. After the 6 weeks the patients are encouraged to resume their pectus breathing and posture exercises and to participate in aerobic sporting activities (soccer, basketball, and swimming). Heavy contact sports (boxing, American football, and ice-hockey) are prohibited until bar removal.

6.3.5 Technique of bar removal (2–4 years after insertion)

The patient undergoes general endotracheal anesthesia with 5–6 cm of PEEP. The patient is positioned supine with both arms abducted at the shoulder. The chest X-rays are reviewed to confirm the position of the stabilizers. Palpation is then performed to see if bar and stabilizer(s) are palpable and close to the old scar. If the bars and stabilizer are not palpable, then "C-arm" fluoroscopy can be used to determine exact site of the hardware. Use the old scars for incision site if at all possible when removing the bar and stabilizer. The bar ends and stabilizers are mobilized and the wire is cut in two places and removed.

When bar and stabilizer have been freed up from the surrounding scar tissue, the inferior wing of the stabilizer is delivered out of the incision followed by the end of the bar and finally the superior wing of the stabilizer. The stabilizer is removed from the bar. The bar is unbent with the bar flippers or multibenders. An orthopedic bone hook is then passed through the hole in the end of the bar and gentle traction is used to slowly extract the bar. The patient is kept on PEEP until the incision is closed.

6.3.5.1 Timing of bar removal

We advise that the pectus bar be left in place for 2–4 years with most patients having their bar(s) out at 3 years. Patients are evaluated on an annual basis and their growth and activity level are monitored. They are encouraged to do their pectus exercises and to participate in aerobic sports. Patients between the ages of 6 and 10 years and 18 years and older often do not grow rapidly, and they tolerate the bar well for up to 4 years. On the other hand, teenagers who undergo a massive growth spurt (15 cm) may require bar removal after 2 years.

6.3.6 Results

6.3.6.1 Demographics

As of December 31, 2007 we have evaluated 1,941 patients with chest wall deformity. The 1,101 patients have undergone pectus repair, 1,015 have undergone primary operations and 86 have undergone redo opera-

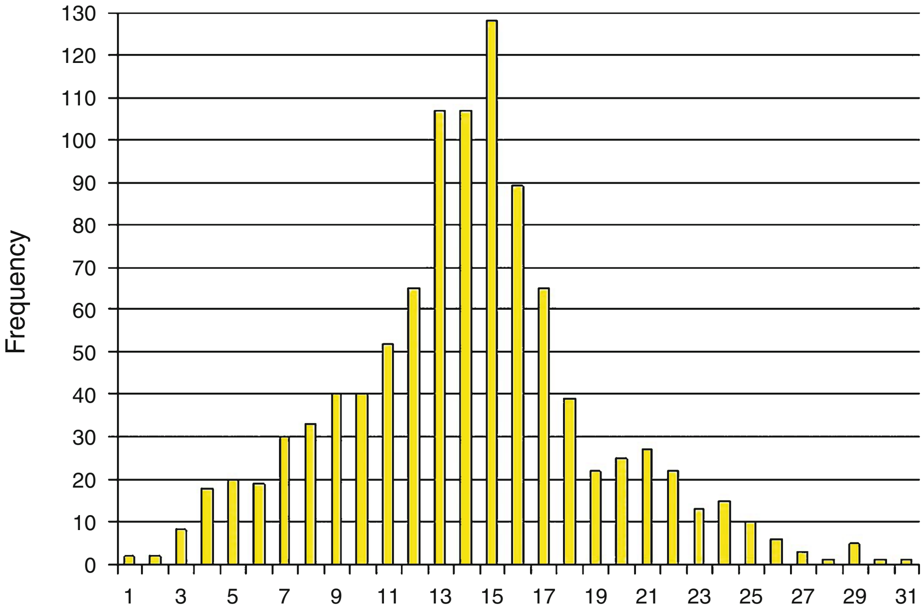

Fig. 16. Graph showing age at primary surgery with a median age of 14 years. Age ranged from 1 to 31 years at our institution

tions. Of the 86 patients undergoing redo procedures, 39 had a failed Ravitch, 44 had a failed Nuss, 1 had both a failed Ravitch and Nuss procedure and 2 had failed Leonard procedure. Of the 1,015 patients having primary repair, 690 patients have had their bar removed. There have been numerous important modifications which have been made both to the surgical technique (e.g., routine use of thoracoscopy) and to the instruments since the origination of this procedure. This has served to facilitate insertion and stabilization of the substernal support bar. These have markedly reduced the risks and complications and have been well documented in recent publications [19].

The male to female ratio in patients undergoing repair was 4:1. The median age was 14 years, with a range of 1–31 years (Fig. 16). The median Haller CT index was 4.6 with a range of 2.4–32.4. Cardiac compression was noted on echocardiography and/or CT scan in 793/889 (89%) patients. Mitral valve prolapse was noted in 132 (15%) patients. Resting pulmonary function testing (PFT) was completed in 900 patients and demonstrated abnormalities in up to 45% of the patients.

6.3.7 Operative procedure, analgesia, and length of stay

A single bar was inserted in 730 (71.9%) patients. Two bars were inserted in 281 (27.7%) patients. Four (0.4%) patients received three bars. Blood loss in most patients was minimal (±10 cc). The median length of stay (LOS) was 5 days with a range of 3–14 days.

6.3.8 Complications

6.3.8.1 Early complications (Fig. 17)

There were no deaths ($n = 0$) nor were there any cardiac perforations ($n = 0$) during the 1,101 primary and secondary repairs performed at our institution. It was common to have pneumothorax after repair but usually it resolved spontaneously. Pneumothorax requiring chest tube drainage occurred in 36 (3.6%) of the primary repairs and required only percutaneous aspiration in 3 (0.3%) primary repairs. In the redo population, 29 (34%) patients required a chest tube placement for evacuation of the pneumothorax with 2 (3%) resolving with aspiration only. Hemothorax requiring drainage but no transfusion occurred after four (0.4%) primary repairs. Three (0.3%) pleural effusions required treatment by either chest tube or aspiration.

In the population of primary repair patients, pericarditis requiring treatment with indomethacin or prednisone occurred following five (0.5%) repairs, with one requiring pericardiocentesis. Pneumonia occurred after 6 (0.6%) repairs, and medication reactions have occurred following 36 (3.6%) repairs. One

• Pneumothorax w/spont. resolution	60.4% ($n = 613$)
• Pneumothorax w/chest tube	3.6% ($n = 36$)
• Horner's syndrome	17.7% ($n = 179$)
• Drug reaction	3.6% ($n = 36$)
• Suture site infection	1.0% ($n = 10$)
• Pneumonia	0.6% ($n = 6$)
• Pericarditis	0.5% ($n = 5$)
• Hemothorax	0.4% ($n = 4$)
• Pleural effusion (requiring drainage)	0.3% ($n = 3$)
• Temporary paralysis	0.1% ($n = 1$)
• Death	0%
• Cardiac perforation	0%

Fig. 17. Early postoperative complications of primary surgical patients

hundred and seventy-nine (17.7%) patients had transient Horner's Syndrome at varying times during the thoracic epidural administration.

6.3.8.2 Late complications (Fig. 18)

Fifty-eight (5.7%) patients have experienced bar displacement, and 43 (4.2%) displacements warranted repositioning. After the introduction of stabilizers, the incidence of bar displacement dropped from 8.9% to 2.3%. There has been only one bar displacement (0.1%) since we combined placing a stabilizer on the left and PDS sutures around the bar and underlying rib on the right. Bar infection occurred in 11 patients (1.1%) requiring early bar removal in 2 (0.2%) patients. Twenty-nine (2.9%) patients had allergies to the bars. These presented in a variety of ways. Several patients gave a history of metal allergy and therefore received a titanium bar, stabilizer and wire suture. The others were diagnosed postoperatively and were treated with antibiotics or steroids. Three of these patients did not respond to medical treatment and required early bar

• Bar displacements	58/1015 (5.7%)
• Requiring revision	43/1015 (4.2%)
• Overcorrection	32/1015 (3.2%)
• Bar allergy	29/1015 (2.9%)
• Wound infection	11/1015 (1.1%)
• Recurrence	8/1015 (0.8%)
• Hemothorax (post-traumatic)	2/1015 (0.2%)
• Skin erosion	1/1015 (0.1%)
• Accidental death	1/1015 (0.1%)
(accidental death 3.5 years post op)	

Fig. 18. Late postoperative complications of primary surgical patients

removal, two of these received titanium bars. The symptoms resolved after removal of the steel bar. Thirty-two (2.9%) developed a moderate overcorrection of their deformity and four (0.4%) developed a true carinatum deformity.

6.3.8.3 Long-term follow-up

Patients are followed at 6 months postoperatively, and then yearly. Long-term assessments classify the post-operative results as excellent, good, fair, or failed.

A result is considered to be excellent if the patient experiences total repair of the pectus excavatum as well as resolution of any associated symptoms. A good result is distinguished by a markedly improved but not totally normal chest wall appearance and resolution of any associated symptoms. A fair result indicates a mild residual pectus excavatum without complete resolution of associated symptoms. And a failed repair indicates a recurrence of the pectus excavatum and associated symptoms and/or the need for another repair after bar removal.

The initial cosmetic and functional results at the time of repair were excellent in 938 (92.4%), patients, good in 74 (7.3%) patients, fair in 1 (0.1%), and failed in 2 (0.2%). Of these patients, 690 (68%) have had their bars out for more than 1 year. The results of the patients who had their bars removed one or more years ago are excellent in 587 (85.4%); good in 75 (10.9%); fair in 9 (1.3%), poor in 7 (1.0%) and failed in 9 (1.3%). There were three patients who did not return (Fig. 19). Patients who did not comply with the exercise program and had their bar removed before puberty had a higher recurrence rate. The length of time the bar was left in situ had a direct effect on the long-term outcome. The longer the bar stayed in, the better the results. The age of the patient also affected the long-term outcome with the best results occurring in the 7–12 and 13–18 year age groups. The long-term results of patients who have had their bar removed before December 31, 2006 show that

• Total number of primary patients	1015
• Total number w/bar removed	690 (64.2%)
– Excellent result	587 (85.4%)
– Good result	75 (10.9%)
– Fair result	9 (1.3%)
– Poor result	7 (1.0%)
– Failed	9 (1.3%)
– No return (bar removed else w)	3 (0.4%)

Fig. 19. Overall results after removal of pectus bar

Number w/bar removed prior to 12/31/06	587
– Excellent result	493 (84.0%)
– Good result	69 (11.8%)
– Fair result	9 (1.5%)
– Poor result	7 (1.2%)
– Failed	9 (1.5%)
– No return	2

Fig. 20. Long-term results in patients having their bar removed before December 31, 2006

we have 587 total patients with and excellent result in 493(84%) patients (Fig. 20).

6.3.9 Conclusion

The minimally invasive procedure provides good to excellent correction of pectus excavatum in over 90% of patients with no rib resection, no sternal osteotomy, minimal blood loss, and rapid return to normal activity. Studies have shown marked improvement in the patient's body image and have also shown slight improvement in pulmonary function [20]. The 1,101 patients managed at our institution have had excellent long-term results and low morbidity.

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