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Minimally invasive surgical repair of pectus excavatum

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The minimally invasive repair of pectus excavatum has become widely accepted. The number of patients presenting for repair has increased dramatically. There have been many technical improvements over 20 years that have made the procedure much safer and more successful. The complications have been identified and preventative measures instituted. The long-term results have shown a 95% good to excellent outcome, and patient satisfaction studies have shown similar results.

The first minimally invasive pectus repair was performed in 1987, and the 10-year experience was published in the *Journal of Pediatric Surgery* in 1998.[1] The experience can, therefore, be divided into the first decade with 42 patients treated at 1 institution and the second decade with several thousand patients treated worldwide.[2-9] In the first decade, the modifications included changing the incision from an anterior chest incision to bilateral thoracic incisions, and the pectus bar was redesigned from a short, soft, square-ended strut to a much longer and stronger steel bar with rounded ends.[1]

In the second decade, a whole host of new features were added to make the procedure safer and more successful. These included routine use of thoracoscopy, the development of new instruments for tunneling and bar rotation, the development of a stabilizer, and placement of pericostal sutures around the bar and underlying ribs to prevent bar displacement.[9-11] The increasing numbers of patients presenting for surgical correction were in part due to an increase in referral by primary care physicians, but also due to self-referral by patients who obtained their information from the Internet.[12-14] This increase in numbers worldwide plus longer follow-up, allowed clarification of age limits[6,8,14-18] and duration of bar placement.[9,19] In addition, complications have been more clearly defined and can be divided into early and late groupings.[20,21] The effects of the early learning experience have been separated from the later experiences.[2,4,13,20-24] More studies comparing cardiopulmonary function before and after surgery are now available[18,25-31] as well as studies comparing quality of life before and after surgery.[32-34] Long-term results after bar removal have confirmed that the excellent results achieved at the time of repair are maintained after bar removal.[7-9,11]

Preoperative considerations

After confirming that the patient's condition is severe enough to fulfill the criteria for surgical correction as outlined by Kelly in this issue of *Seminars in Pediatric Surgery*, several other factors need to be considered.

Age

The minimally invasive repair has been performed successfully on patients from 1 year to over 50 years.[6,8,10,14-19] The ideal age is just before puberty because at that age the chest is still very malleable, the support bar is in place during the pubertal growth spurt, the recovery time is short, and the incidence of recurrence is low.[9,19] Patients less than 8 years of age also have an excellent result and short recovery time,

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but because the support bar is removed before the pubertal growth spurt, there is potential for recurrence.[9,35] However, if a young patient has significant cardiac and/or pulmonary compression, then an early repair is justified. The family needs to be informed that the patient may require a second bar placement either at the time of removal of the first bar when a longer bar may be inserted by using a "chest tube switch technique" or inserting it later if a recurrence develops during puberty, which occurs in approximately 5% of patients. During the first decade, it was thought that the minimally invasive procedure was only useful in prepubertal patients, but experience has shown that postpubertal patients tolerate the procedure well, and excellent results have been reported in patients in their 30s and 40s.[9,15-19] The older patients require two or more bars in more than 50% of the cases.[9]

Phenotype

The ideal chest configurations for the minimally invasive repair are the diffuse "saucer shape," localized "cup shape," and symmetric funnel shape.[9] Patients who have very steep cup-shaped depressions and patients with severe deep asymmetric "grand canyon type" depressions are more of a challenge and often require two bars. Park and others have suggested using an asymmetric bar in these patients.[6,7] In patients where the depression involves mostly the upper chest, care needs to be taken not to place the bar too high as it will interfere with the axilla and its vital structures. Patients who have mixed excavatum/carinatum deformities may have residual protrusion of the carinatum post bar placement, especially if there is severe sternal torsion. Older patients have a higher incidence of sternal torsion and mixed deformities, which may be a good reason to perform the repair before puberty.[16,36,37] Patients who have a "pouter pigeon" deformity with anterior displacement of the manubrium and posterior displacement of the gladiolus develop increased protrusion of the manubrium when the gladiolus is elevated with a substernal bar. We, therefore, do not recommend the minimally invasive procedure in this category of patients.

Pre-operative preparation

If the patient has a history of nickel allergy, which occurs in 2% of the population, then a titanium bar should be used.[38] This requires advanced planning as the bar needs to be ordered from the manufacturer before surgery (Biomet Microfixation, Jacksonville, FL). In order that the bar can be prebent and polished, to prevent tissue adherence, the manufacturer will need to know the length of the bar required and a copy of the CT scan at the insertion site. All the usual precautions for insertion of a foreign body into a patient need to be meticulously adhered to. Pain management is discussed with the patient and family, and the risks and complications of epidural analgesia need to be reviewed as well.

Positioning the patient

The standard position is supine with both arms abducted at the shoulders to approximately 70°, taking care to protect the patient from brachial plexus injury. Alternative methods include elevating the torso on a mattress and extending the arms posteriorly.[22,39] This position allows insertion of the thoracoscope superior to the incision site. It has the disadvantage of over-extending the chest during the surgery. Another alternative position is to flex the left shoulder and elbow anteriorly, adjacent to the head,[40] but there have been anecdotal reports of brachial plexus injury with this position.

Thoracoscopy

Thoracoscopy has become a routine part of the minimally invasive procedure.[11,41,42] Most surgeons use right-sided thoracoscopy,[43] others prefer left-sided thoracoscopy,[40,44] some use bilateral thoracoscopy,[45,46] and some insert the scope and introducer through the same thoracostomy sites.[47] In patients with extremely deep depressions, it may be necessary to use bilateral thoracoscopy because the heart is not only compressed, but is also displaced to the left, which impedes visibility from the right. Insertion of the trocar from the left when the heart is displaced in that direction requires great caution. The trocar is usually inserted inferior to the incision sites, but it can be inserted through the incision site[13,40] or even superior to the incision site when the arm is extended posteriorly.[22,39] The trocar insertion site affects visibility and the inferior insertion site allows for good visibility not only during the tunneling but also for suture placement during bar stabilization. We use blunt instrumentation for trocar insertion and direct the trocar in a superior direction so as to avoid the liver and diaphragm.[43]

The introducer tip should always be kept in view through the thoracoscope during the mediastinal tunneling. If the tip cannot be visualized because the depression is too deep, then the scope may be inserted from the opposite side or a more superior tunnel should be created first where the depression is not so deep. A 30° or flexible scope is helpful in this situation.

The CO_2 insufflation pressure should be kept as low as possible and usually a pressure of 5 mm Hg is sufficient to keep the lungs out of the operative field. When two bars are being inserted, there will be more leakage, requiring a higher flow rate to keep the pressure up.

Skin incision site

During the early days of the procedure, the anterior thoracic incision used for open repairs was also used for the minimally invasive procedure. However, this incision resulted in keloid formation because of tension on the wound, and it

was difficult to place the bar ends into the subcutaneous pouch without extending it all the way across the chest. A decision was therefore made to insert the bar through two small lateral thoracic incisions.[1] Transverse lateral thoracic incisions have the advantage of providing good access to the thoracostomy entry and exit sites, run parallel to the lines of tension (Langers Lines), rarely cause keloid formation, and require minimal subcutaneous dissection. Vertical incisions in the mid- or posterior axillary lines give poor access to the anterior chest wall and tend to cause keloid formation. When placing two or more bars, making a separate incision for each bar facilitates bar stabilization and bar removal after 3 years. In mature female patients, the incisions should be placed in the inframammary crease between the 6- and 9-o'clock position and extended as necessary. The inframammary incisions give excellent access to the anterior chest wall, even allowing insertion of two bars, and give an excellent cosmetic result since the incisions virtually disappear.

Tunneling

The thoracic entry and exit sites should be placed close to the sternum to prevent disruption of the intercostal muscles. Ideally, the tunnel should pass right under the deepest point of the depression. If the deepest point of the deformity is inferior to the body of the sternum, then the patient requires two bars: one under the sternum and one under the deepest point of the depression. The introducer tip should always be kept in view during the tunneling. If the depression is too deep for this to occur, then the first tunnel should be created more superiorly, leaving the introducer in place to elevate the sternum. Alternatively, the sternum can be elevated by using the suction cup[44] or lifting it with a towel clamp or heavy suture.

An extra-pleural approach has been advocated by Schaarschmidt to prevent pleural and pericardial reaction with good preliminary results.[45] This is technically more difficult, and the internal mammary vessels are at increased risk of being injured.

Sternal elevation

When the introducer is in position across the mediastinum, it is lifted in an anterior direction to pull the sternum and anterior chest wall out of their depressed position, thereby correcting the pectus excavatum. Repeating this lifting maneuver several times loosens up the anterior chest wall, prevents the substernal trauma and intercostal muscle injury caused by bar rotation, and minimizes the pressure on the bar, which decreases the risk of bar displacement. The pectus excavatum should be completely corrected before removing the introducer.

Bar stabilization

Bar stabilization is essential for a successful outcome. When the minimally invasive technique was first developed, bar stabilization was attempted by creating a muscular pocket.[1] This technique resulted in a 15% bar displacement rate. Subsequently, a stabilizer or foot plate was developed and attached to the bar to give it more stability.[11] Initially, the stabilizer was only held in position with fascial sutures, but it frequently became detached from the bar and so it was decided to lash the stabilizer to the bar with wire sutures. However, even with a stabilizer attached, some patients dislodge their bar during the first 3 weeks before scar tissue can be laid down and there is therefore a need for additional support during those first few weeks after surgery. Hebra and coworkers[10] were the first to advocate placing a suture around the bar and underlying ribs and called it "third point fixation."[10] They advocated placing the suture adjacent to the sternum through a small stab wound. Most surgeons now use the lateral thoracic incision to place sutures around the bar and rib under thoracoscopic control.[8,9,48] Some centers use wire instead of absorbable sutures.[6,23,36,41] This does increase the risk of injury to the underlying lung, especially if the wire fractures.[37] A new absorbable stabilizer which is slowly reabsorbed over a 6- to 12-month timeframe has recently become available. There have been no complications reported with its use at this time.

Number of bars

Initially the procedure was done only on young patients and so only one bar was necessary.[1] However, now that the procedure is being used more commonly in postpubertal patients, it has been noted by numerous investigators that two bars give better and more stable results.[1,18,48] Patients with Marfan syndrome, asymmetric "grand canyon"-type deformities, and wide saucer-shaped deformities also usually require two bars.[1,7,9,36] A second bar should be inserted if the repair is suboptimal after insertion of one bar. On the operating table, the correction always looks better than it does when the patient resumes normal posture because the normal thoracic lordosis is eliminated on the operating table. I have never regretted placing a second bar but have often regretted placing only one.

Bar and chest configuration

Many of the patients who have had to have a re-operation have come to us because they were initially undercorrected. It is important to slightly overcorrect the deformity to prevent "buckling" of the anterior chest wall and to decrease the risk of recurrence. The bar should therefore have a semicircular shape with only a 2- to 4-cm flat section in the middle to support the sternum. The thoracostomy entry and

exit sites into and out of the chest should be medial to the top of the pectus ridge on each side. A bar that is only bent at each end ("table top configuration") will give insufficient correction and may allow the lung to herniate between the bar and anterior chest wall. The bar should not be too tight on the sides of the chest because it will cause painful rib and muscle erosion and the patient will outgrow the bar too soon, necessitating early bar removal.[7,24] In asymmetric patients, Park and others have recommended using an asymmetric bar, which gives more lift on the side of the asymmetric deformity.[6,8]

Re-operation

Re-operations on failed previous repairs have been successfully accomplished in 44 previous Nuss repairs, 40 previous Ravitch repairs, and 2 previous Leonard repairs. Thoracoscopy is particularly important in this group of patients as they usually require lysis of adhesions before the tunneling can commence, and that requires an additional port placement.[45] A postoperative chest tube is helpful in managing the inevitable lung leak and oozing that follows the lysis of pulmonary adhesions. The failed open procedures fall into two categories: those in whom there was a "simple cave-in" in after the open repair which are easily corrected with a substernal bar, and those in whom there is diffuse osteochondrodystrophy who are not amenable to correction with a substernal bar because of excessive calcification and rigidity of the chest wall. In this group with severe recurrence, acquired asphyxiating chondrodystrophy, and a rigid chest wall, we have had 2 arrhythmic arrests thought to be due to pressure of the introducer on the heart. Both patients were resuscitated, but in 1 patient sternotomy was required as the rigid calcified chest wall did not allow external cardiac compression. Complications are higher in the re-operation group of patients as 44% required chest tubes, 8% had hemothorax without requiring transfusion, 8% had pleural effusion requiring drainage, and 2 patients had temporary arrhythmic arrest as mentioned above. Bar displacement occurred in 9%. The results are excellent in 70%, good in 28%, and fair in 2%, which is slightly less satisfactory than the primary repair group.[36,49,50]

Prior thoracic surgery and concomitant open or thoracoscopic intrathoracic procedures have been successfully performed in conjunction with the minimally invasive pectus excavatum repair.[51]

Pain management

Two years ago, we adopted the preemptive pain management protocol used in other specialties. The purpose of this protocol is to prevent the pain cascade from being triggered in the first place, rather than reacting to it after the fact. We also decided to deal with the patients' anxiety in a proactive manner, since fear is well known to decrease the pain threshold. All patients now receive lorazepam on the night before the surgery so that they will arrive at the hospital well rested and less anxious. In the presurgical holding area, they receive p.o. midazolam 45 minutes before being taken to the operating room. The younger patients are anesthetized and intubated before the epidural catheter is inserted, and the older patients have the epidural inserted under moderate sedation and are then anesthetized. While the patients are being prepared for surgery, a dose of IV ketorolac is given, and ketorolac is continued every 6 hours until 1 day after the epidural is removed, on day 3 or 4. While the patient is receiving the ketorolac, the IV fluids are kept at maintenance level to flush the kidneys, and famotidine is given to prevent gastritis and gastrointestinal ulceration. In addition, blood urea and creatinine levels are checked on days 1 and 3 postoperation. Epidural infusion of fentanyl and bupivacaine is started during induction of anesthesia and continued until day 3 or 4 postoperation. Patients also receive low-dose diazepam for muscle relaxation and anxiolysis. Morphine is used for breakthrough pain and has become rarely necessary since starting this pain protocol. We also start stool softeners and laxatives prophylactically on day 1 to prevent constipation. Since instituting this protocol, there has been a dramatic improvement in postoperative pain control, appetite, and patients' ability to ambulate starting on day 1. Patients are discharged on day 4 or 5 with oxycodone and ibuprofen for pain and diazepam and robaxin for muscle relaxation.

Patients may return to school whenever they are strong enough, which varies with age. Prepubertal children recover quicker and are usually ready to return in 2 weeks, whereas postpubertal patients require 3 weeks. All patients are re-

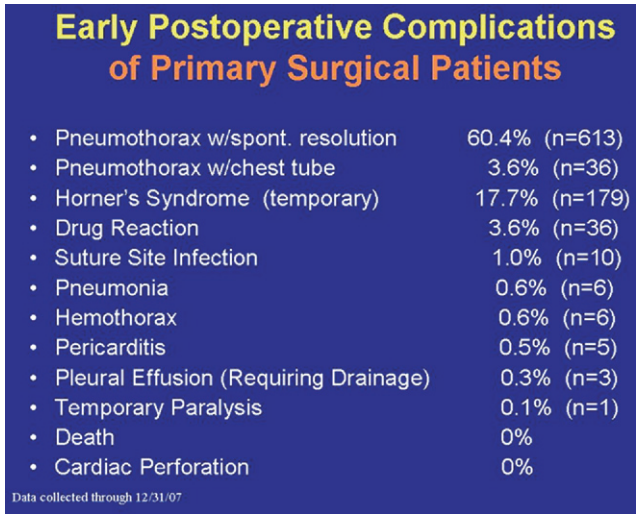

Figure 1 Early post operative complications encountered in 1,015 primary operations performed between 1987 and 2008. The Horner's syndrome was a side effect of epidural analgesia and resolved spontaneously in all cases. (Color version of figure is available online.)

stricted from participating in sporting activities for a minimum of 6 weeks and competitive sports for 8 to 10 weeks.

Early postoperative complications (Figure 1)

Early complications have been markedly reduced by meticulous attention to fitness for surgery, surgical technique, bar stabilization, evacuation of the pneumothorax, incentive spirometry, and prophylactic antibiotics. Many centers have reported marked improvement in their complication rate after the early learning experience.[11,13,20,21,24]

The most common "complication" is an insignificant residual pneumothorax secondary to CO_2 insufflation at thoracoscopy, which resolves spontaneously. A chest tube was inserted in 3% of our patients usually because the CO_2 was not adequately removed or because there was a leak in the system before removal of the trocar and the surgeon elected to leave a chest tube rather than take the risk of having to insert one later. There should not be a lung leak in a primary pectus repair, especially if the trocar is inserted after first creating a blunt thoracostomy. On the other hand, "re-do" operations with lysis of pulmonary adhesions frequently require a postoperative chest tube.[45,49] Pneumonia is very rare in these young patients (0.6%), but postoperative incentive spirometry is vigorously encouraged, and all patients are given prophylactic antibiotics (cefazolin) for 5 days. Wound and/or bar infection can be prevented if all the precautions for foreign body insertion are meticulously adhered to and occurred in less than 1% of patients.[52-54] Infection requires vigorous treatment consisting of wound drainage, cultures and appropriate intravenous antibiotics, followed by long-term oral antibiotics (sulfamethoxazole/trimethoprim). Treatment is usually effective in saving the bar if it is continued until the ESR and CRP have returned to normal levels.[52-54] There have been reports of an increased infection rate on the side with the stabilizer,[55] but that has not been our experience in more than 1000 cases.

Pericarditis occurred in 0.4% of our patients.[38,56,57] The etiology is unclear. It may be due to nickel allergy, pericardial trauma, or postcardiomyotomy syndrome. These patients present with persistent central chest pain, malaise, lethargy, and a pericardial friction rub. If an echocardiogram confirms that there is pericardial fluid present, then the patient should be treated with a short course of prednisone. A pleural effusion that lasts more than 4 days may also be due to nickel allergy and should be treated similarly after aspirating fluid for culture. If symptoms recur after the prednisone has been discontinued, then the patient should be tested for nickel allergy (T.R.U.E. patch; Allerderm, Phoenix, AZ). If positive, the options are to give low-dose prednisone on alternative days until the ESR and CRP return to normal or to replace the steel bar with a titanium bar.

Cardiac perforation has occurred in several centers during the early learning experience and before thoracoscopy was widely available.[6,58-60] Reviewing the preop CT scan to determine the position of the heart and its relationship to the sternum is very helpful in planning the procedure, especially in patients with severe asymmetry and sternal torsion. If it appears that the heart is severely compressed, then elevating the sternum with a hook or suction cup during the tunneling greatly minimizes the risk of injury. In addition, by first tunneling one or two intercostal spaces superior to the deepest point and leaving the introducer in place to keep the sternum elevated while creating the second tunnel, also minimizes the risk of pericardial or cardiac injury. The tip of the introducer should always be kept in sight. Good visibility with the thoracoscope in place is essential and, if necessary, bilateral thoracoscopy should be used.

It should be noted that, with a substernal bar in place, cardioversion requires placement of the paddles in an anterior–posterior position so that the current will be conducted through the heart. If the paddles are placed anterior and lateral, then the current will simply be conducted along the bar and not through the heart.[59]

Late complications (Figure 2)

Bar displacement has been the biggest late challenge. The initial bar displacement rate was 15%. After the introduction of stabilizers, it dropped to 5%, and with the addition of pericostal sutures placed around the bar and underlying ribs, it dropped to less than 1%.[9,20] Our standard procedure is to place a wired stabilizer on the left and multiple double-stranded "0" PDS pericostal sutures around the bar and underlying ribs on the right. If feasible, we also place pericostal

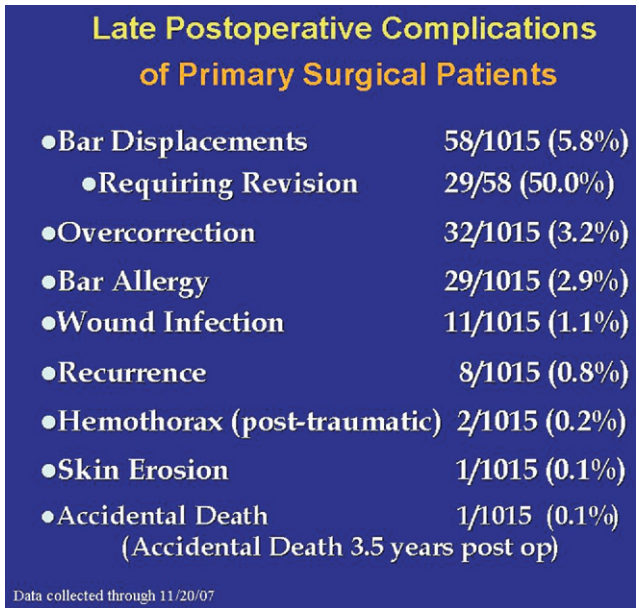

Figure 2 Late post operative complications encountered in 1,015 primary operations performed between 1987 to 2008. The "accidental death" occurred as a result of trauma unrelated to the pectus excavatum. (Color version of figure is available online.)

sutures on the left. In our practice, only 50% of all the displacements required revision. If the displacement is less than 20% and the repair remains excellent, it can be observed. If there is no further progression, then surgical revision is not usually required.

Nickel allergy, which is present in 2%[38] of the population, may manifest early with pericarditis or persistent pleural effusion, but may also occur late with erythema of the anterior chest wall or inflammation and drainage at the incision sites. The inflammation and drainage may resemble a chronic infection, but cultures are negative and testing for nickel allergy will give a positive result. Treatment consists of local wound care and a short trial of prednisone. If the patient responds, then low-dose alternate day prednisone until the ESR and CRP are back to normal will usually resolve the problem. If the patient responds to the steroid therapy, the bar can be left in place until it is time for removal. If the patient does not respond to treatment, then the steel bar needs to be replaced with a titanium bar.[38]

Overcorrection, resulting in pectus carinatum, has occurred in 0.4% of our patients. These patients all suffered from Marfan syndrome and had very deep cup-shaped deformities. Early bar removal was successful in one patient, and two required an external pressure brace. Others have reported on carinatum developing, especially in patients with asymmetry and a twisted sternum.[23]

Undercorrection not only predisposes the patients to increased risk of recurrence but also results in abnormal ridges developing adjacent to the sternum because there is not enough space. The cartilaginous portion of the rib will buckle under the pressure.

Persistent pain may be due to bar displacement, stabilizer dislocation, bar too tight, bar too long, sternal or rib erosion, infection, or allergy. An anterior and lateral chest x-ray, complete blood count, ESR, CRP, and T.R.U.E. patch for allergy will identify the cause and allow appropriate treatment.

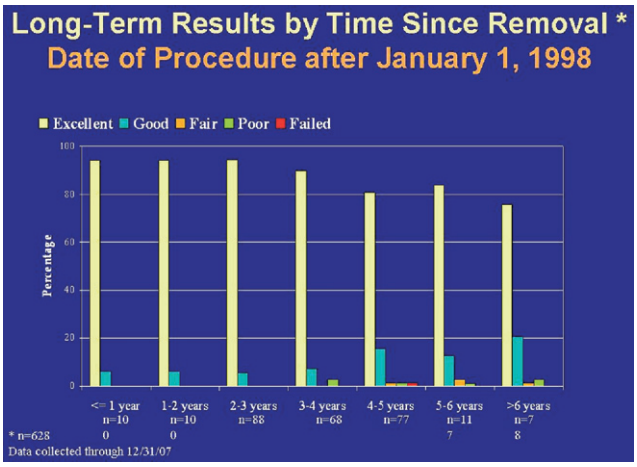

Figure 3 Results in 628 patients more than one year after bar removal. (Color version of figure is available online.)

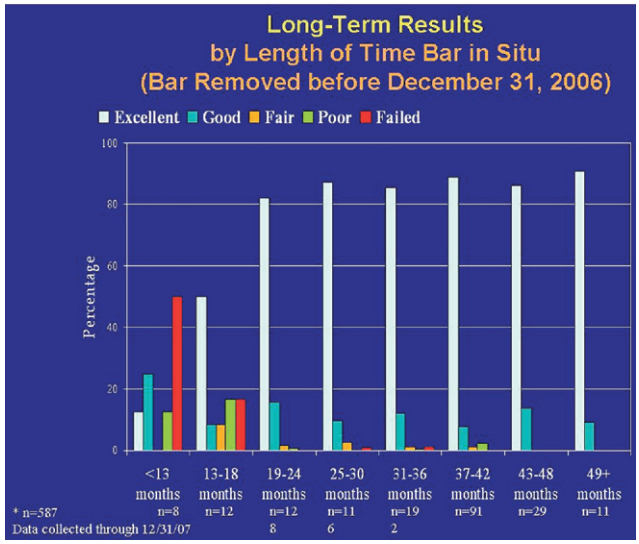

Figure 4 Results in 587 patients more than one year after bar removal showing inverse relationship to duration of bar placement. (Color version of figure is available online.)

An accidental death unrelated to the pectus surgery occurred when a patient fell off an eighth story balcony 3.5 years after his pectus repair.

Results

Long-term results in 628 primary repair patients who are more than 1 year post bar removal are excellent in 540 patients (86.0%), good in 65 patients(10.3%), fair in 15 patients (2.4%), and failed in 8 patients (1.3%) (Figure 3).

Similar results have been reported by other centers.[5-8,24] Long-term results have shown that the bar should remain in situ for 2 to 4 years (Figure 4).

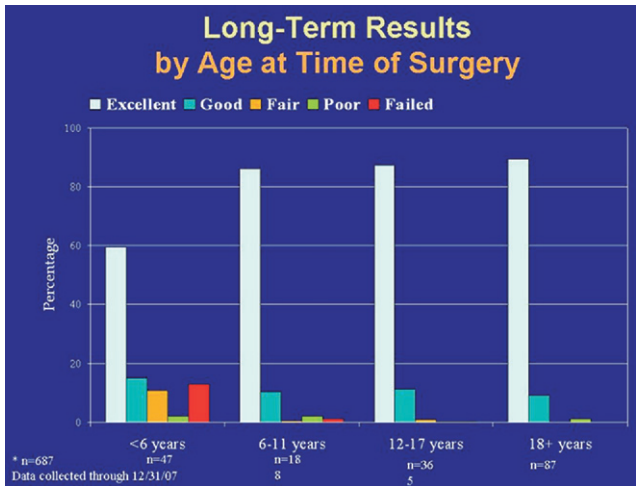

Figure 5 Results in 687 patients showing less satisfactory longer-term outcome in patients less than 6 years of age. (Color version of figure is available online.)

If the bar is removed before 2 years, the recurrence rate increases inversely with the length of time the bar remains in situ.

The age at the time of repair affects recurrence rate (Figure 5). If the bar is removed before puberty, there is increased risk of recurrence.[9]

All patients are encouraged to exercise regularly starting 6 to 8 weeks postoperatively. We believe that patients who exercise regularly are more likely to maintain their excellent result than patients who are sedentary and rarely expand their chest to full capacity.

Postop cardiopulmonary function studies have shown good improvement in some studies and less so in other studies. Several studies have shown significant improvement in pulmonary function postoperatively,[25-27,30,31] whereas other studies have shown no significant change.[18] The reasons for this discrepancy are multifactorial and include the size of the cohort being studied, the duration of the study, the severity of the pectus excavatum, whether the studies were done during exercise or at rest, etc. In our series of more than 900 cases, the preoperative resting pulmonary studies showed a marked shift to the left and a significant correction postoperatively[26,31] (Figures 6 and 7).

Postoperative cardiac studies have shown an increase in cardiac filling and stroke volume postoperatively.[28-31] There is a discrepancy between the overwhelming number of anecdotal reports by the patients versus the results of cardiac and pulmonary studies. Surgeons regularly hear, "I never realized how incapacitated I was until I had my pectus corrected" or "I played basketball before the surgery but required frequent breaks. Now I am the fastest on my team and I can play an entire game without stopping," or similar comments.

Quality-of-life studies and overall patient satisfaction studies have shown a significant improvement in patient self-esteem and level of satisfaction following the minimal invasive repair.[8,14,32-34]

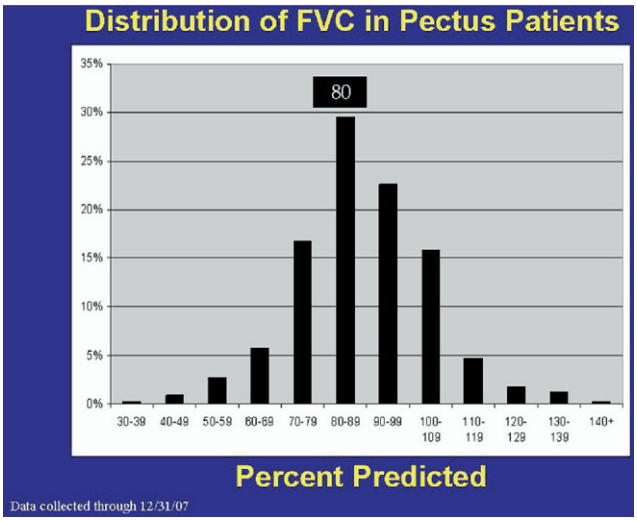

Figure 6 Pre-op resting pulmonary function studies in 900 pectus excavatum patients before, surgery, showing FVC shifted to the left. The graph should peak at 100 percent of predicted value and not at 80. (Color version of figure is available online.)

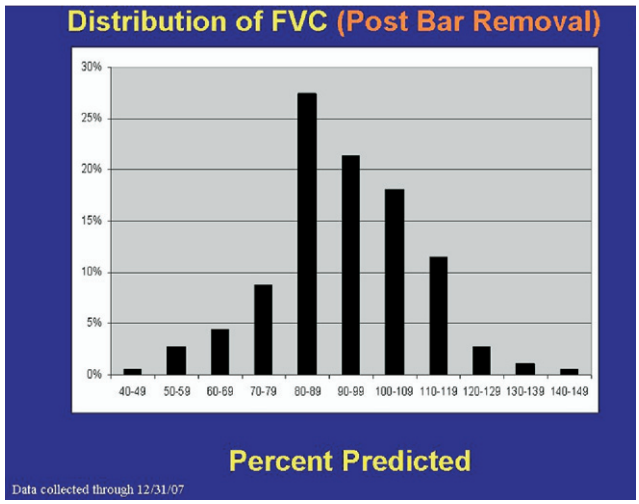

Figure 7 Post-op resting pulmonary function studies showing that the FVC has shifted towards normal. (Color version of figure is available online.)

Bar removal (see Figure 4)

The bar(s) should remain in the chest for 2 to 4 years after pectus repair. Most patients tolerate the bar well for 3 years and are able to participate in competitive sports. There have been a few patients who have kept their bar in situ for 4 or more years without any problems. If a patient grows more than 6 inches(13 cm) after bar insertion and becomes symptomatic with lateral chest pain, then he needs to be evaluated to see whether early bar removal is required.

Bar removal is accomplished under general anesthesia with positive pressure ventilation and 5 to 6 cm of PEEP to prevent pneumothorax. Both sides of the bar should be mobilized, and the bar should be unbent by using either the bar flippers or Multibenders (Biomet Microfixation, Jacksonville, FL).[45,61,62] After straightening, the bar is removed very slowly while monitoring the EKG and all the other vital signs. A postoperative chest radiograph is done routinely to check for pneumothorax. Our complication rate for bar removal in 700 patients was 3 pneumothoraxes requiring aspiration and 1 wound infection. Elsewhere, there have been isolated reports of major complications, including one cardiac arrest and a pulmonary hemorrhage requiring thoracotomy.[63]

Conclusion

In the 20 years since the first minimally invasive pectus excavatum repair was performed, numerous modifications have made the procedure safer and more successful. As a result there has been a dramatic increase in the number of

patients seeking surgical correction. Recent studies have confirmed a reduction in the complication rate after the early learning experience, an improvement in excellent results, and 95% overall patient satisfaction rate.[14,41]

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