Indications and Technique of Nuss Procedure for Pectus Excavatum

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KEYWORDS

• Pectus excavatum • Thoracoscopy • Nickel allergy
• Nuss procedure

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

In the second decade, many new features were added to make the procedure safer and more successful. These features included the routine use of thoracoscopy; the development of completely new instruments specifically designed for tunneling, bar rotation, and bar bending; the development of a stabilizer; and the placement of pericostal sutures around the bar and underlying ribs to prevent bar displacement.[9–12] The increase in the number of patients presenting for surgical correction was not only because of an increase in referral by primary care physicians but also because of self-referral by patients who obtained their information from the Internet.[12–14] This increase in numbers worldwide, combined with longer follow-up, allowed clarification of age limits, indications for surgery, and duration of bar placement.[6,8,9,12,14–19] In addition, complications have been more clearly defined and are divided into early and late groupings.[20–22] The effects of the early learning experience have been separated from those of the later experiences.[2,4,13,20–26] More studies comparing cardiopulmonary function[18,27–33] and quality of life[34–37] before and after surgery are now available. Long-term results after bar removal have confirmed that the excellent results achieved at the time of repair are maintained after bar removal.[7–9,11,12,38]

CLINICAL FEATURES

Pectus excavatum (PE) may be present at birth, but in the authors' series of more than 2000 patients, most presented during the pubertal growth spurt, of which 80% were boys (**Table 1**).[38] Associated scoliosis occurs in 20% to 30% of the patients,[38,39] and connective tissue disorders such as Marfan syndrome, Marfanoid features and Ehlers-Danlos syndrome occur in up to 20%.[38]

Morphology

The deformity most frequently involves the lower sternum and chest wall. Focal or cup-shaped depressions are the most common type; broad, shallow, saucer-shaped deformities are the second most frequent; a long furrow or trench, which is usually asymmetrical (Grand Canyon

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Thorac Surg Clin 20 (2010) 583–597
doi:10.1016/j.thorsurg.2010.07.002
1547-4127/10/$ – see front matter

Table 1
Medical history of 327 patients studied

Condition	Number	Percentage
Exercise intolerance	211	64.5
Lack of endurance	205	62.7
Shortness of breath	203	62.1
Chest pain with exercise	167	51.1
Family history of PE	140	42.8
Chest pain without exercise	104	31.8
Asthma	70	21.4
Scoliosis	69	21.1
Cardiac abnormalities	65	19.9
Frequent or prolonged URI	44	13.5
Palpitations	37	11.3
Pneumonia	28	8.6
Fainting/dizziness	27	8.3
Marfan syndrome	15	4.6
Family history of PC	13	4.0
Ehlers-Danlos syndrome	9	2.8
Family history of Marfan syndrome	8	2.4
Patient adopted	4	1.2
Patient has identical twin	3	0.9
Family history of Ehlers-Danlos syndrome	2	0.6
Sprengel deformity	2	0.6

Abbreviations: PC, pectus carinatum; URI, upper respiratory infection.

From Kelly RE Jr. Pectus excavatum: historical background, clinical picture, preoperative evaluation and criteria for operation. Semin Pediatr Surg 2008; 17(3):210; with permission.

type), is the third most common; and mixed carinatum and excavatum deformities occur in 5%.[40]

Radiographic Evaluation

Because the morphology varies, preoperative imaging for anatomic assessment and documentation of dimensions of the chest are important. A routine chest radiograph is used in some centers[41] because it is inexpensive, readily available, and allows measurement of the indices of severity. The radiograph is also helpful in recurrent PE because it shows the extent of abnormal calcification of cartilages.

A computed tomographic (CT) scan of the chest without contrast gives a clearer picture of the deformity and the bony and cartilaginous skeleton in 3 dimensions and allows calculation of the CT index. The 3-dimensional reconstruction is useful in determining the number of bars that may be necessary, especially in diffuse deformities, which extend up toward the clavicles. Cartilaginous deformity is poorly seen on a chest radiograph but well visualized on a CT scan. Likewise, cardiac and pulmonary compressions as well as the relationship of the sternum to the compressed heart are much better visualized on the CT scan than on a chest radiograph. CT also helps better to see abnormal calcification of cartilages in a recurrent previous open (Ravitch) operation.[42] Review of the CT scan with the patient and parents before surgery helps to communicate the extent of deformity and to form expectations for the hospital course and final outcome. A method for measuring asymmetry and cephalad extension of the depression by CT scan has been published.[43]

Magnetic resonance imaging (MRI) may be used instead of CT to reduce radiation exposure, especially in children who are old enough to cooperate and do not require sedation or general anesthesia for MRI.

Daunt and colleagues[44] examined normal children to obtain values for the pectus index (2–2.3 in normal individuals). In 557 patients, it was found that the 0- to 2-year age group had a significantly smaller mean Haller (pectus) index than older children. In addition, girls had significantly greater pectus index values than boys in the 0- to 6-year and 12- to 18-year age groups. Following surgical correction, Kilda and colleagues[45] observed that the pectus index increased by 0.45 ± 0.49. Statistically significant index differences before and after surgery were not detected in 88 children when the preoperation pectus index was less than 3.12 ($P = .098$). The investigators recommended indications for surgical treatment based on improvement in values for several radiographic indices after operation. Kilda and colleagues state that the commonly used pectus index should be greater than 3.1.

Exercise Limitation

Many patients with PE have a perceived limitation of exercise ability.[30,37] Investigations of exercise ability have yielded mixed results. Over more than 50 years, there have been dozens of studies of cardiac or pulmonary function in PE. In 2006, Malek and associates[30] reported a meta-analysis based on a computer-assisted search of the literature. The investigators concluded that surgical repair improved cardiovascular function. Sigalet and colleagues[32] studied patients pre-and postrepair and reported significant improvement in cardiac stroke volume, cardiac output, forced expiratory volume in the first second of expiration (FEV_1), total lung capacity (TLC), TLC (percentage expected), diffusing lung capacity, maximum oxygen consumption (Vo_2max), respiratory quotient, and O_2 pulse (percentage predicted).

Pulmonary Function Studies

Efforts to elicit the cause of exercise intolerance have led to studies of pulmonary function at rest, including spirometry and plethysmography. Results of spirometry in patients with PE are usually 10% to 20% less than the expected average for the population. Plethysmography shows that lung volumes are similarly decreased (**Fig. 1**).[46–48] At the authors' institution, in 855 patients with PE presenting for surgical treatment, the mean forced vital capacity (FVC) was only 77% instead of the 100% predicted value, the mean FEV_1 was 83%, and the forced expiratory flow, midexpiratory phase ($FEF_{25\%-75\%}$) was 73%. In the same group, 26% of the patients had an FVC in the abnormal category, less than the 80% predicted value, and for FEV_1 and $FEF_{25\%-75\%}$, the number in the abnormal range was even higher, with 32% and 45%, respectively, whereas in the normal distribution, only 16% of the patients should have less than the 80% predicted value ($P<.001$). In 327 patients enrolled in a multicenter study of PE, FVC for patients aged 8 to 21 years was a mean 90% of the predicted value; FEV_1, 89%; and FEF $_{25\%-75\%}$, 85%. These decreases are statistically and clinically significant ($P<.001$) (see **Fig. 1**).[12,28,33] In a study of patients with recurrent PE, the predicted values showed significant restrictive disease in more than half of the patients.[49]

Cardiology Evaluation

Cardiological evaluation is important because a significant number of patients have findings of right atrial and ventricular compression, mitral valve prolapse, and rhythm abnormalities. Also, because many patients with PE have exercise-related symptoms, including chest pain, it is useful to assure normal heart functioning when planning a major thoracic operation. Mitral valve prolapse was present in 17% of the patients in the authors' series and in up to 65% of those in other series, as opposed to only 1% in the normal pediatric population.[50–52] Mitral valve prolapse as a direct consequence of compression is suggested by CT scan[53] and confirmed by its resolution in half of the surgically treated cases. Dysrhythmias, including first-degree heart block, right bundle branch block, or Wolff-Parkinson-White syndrome, were present in 16% of the authors' patients.[54]

The hemodynamic effects of PE have been the subject of numerous reports and much controversy. The amount of right atrial and ventricular compressions varies with the overall shape of the chest.[55] These effects were reported first several years ago, but more recent imaging and exercise/work/oxygen uptake techniques are clarifying previous findings. Using angiography, Garusi and D'Ettorre[56] demonstrated displacement of the heart to the left side with a sternal imprint on the anterior wall of the right ventricle.[50,56] This finding is now easily verified noninvasively by CT or MRI.

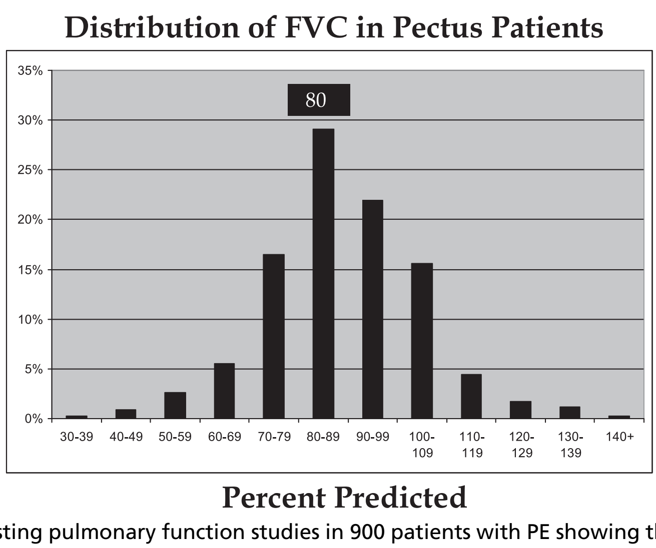

Fig. 1. Preoperative resting pulmonary function studies in 900 patients with PE showing the peak for FVC shifted to the left. The graph should peak at 100% of predicted value and not at 80%. (*From* Nuss D. Minimally invasive surgical repair of pectus excavatum. Semin Pediatr Surg 2008;17(3):215; with permission.)

In 1962, Bevegard[57] found that work capacity was related to the severity of the pectus depression. Evaluating 16 patients via right heart catheterization, he found that patients with a 20% or greater decrease in physical work capacity had a shorter distance from the sternum to the vertebrae. The decrease in stroke volume on changing from a supine to sitting posture at rest was similar to that of normal subjects (40%). The increase in stroke volume from rest to exercise was only 18.5%, much less than the 51% increase in normal subjects ($P<.001$). Thus a measured lower work capacity was found at any given heart rate while sitting.[50,57] Beseir and colleagues[58] performed cardiac catheterization on 6 patients and showed that stroke volume was 31% lower and cardiac output was 28% lower during upright and supine exercises. Postoperation, 3 patients had a 38% increase in the cardiac index, entirely because of an increase in stroke volume.[58]

The effect of mechanical compression by the sternum on the heart was demonstrated by Heitzer and Wollschlager[59] in a patient with a severe deformity and ischemic changes correlated by catheterization, CT scan, and electrocardiogram (ECG).[58] The occurrence of cor pulmonale and chronic respiratory acidosis is rare, but these conditions have been reported in a patient with PE in whom no other cause was uncovered after an extensive diagnostic workup.[60]

The narrower the chest in anteroposterior direction the more the heart is apt to be squeezed between the sternum and spine and the stroke volume diminished, as has been demonstrated by direct cardiac catheterization, oxygen saturation studies, CT scanning, and echocardiogram.[30,32,61] Improvement after operation has been demonstrated.[30,32,61] One of the reasons for persisting controversy concerning cardiac effects of PE until recently is the failure to objectively measure the severity of the chest depression and correlate it to the amount of cardiac compression. Kinuya and colleagues,[29] Sigalet and colleagues,[32] Coln and colleagues[31] and other researchers have clearly demonstrated the relief of cardiac compression.

In summary, compression of the right side of the heart, leading to a diminished stroke volume, combined with a modest lung restriction, leads to a diminished cardiopulmonary capacity in severe cases.

Body Image

Concern about the appearance of the chest prompts many patients to seek repair. A large percentage of patients with PE are extremely self-conscious about their chests; these patients withdraw from social and sports activities and become depressed and even suicidal. Children and adolescents with visible physical differences are at risk for developing body image issues and interpersonal difficulties because they are teased by their peers, which further aggravates the problem.[62] Nevertheless, patients with PE are often dismissed by pediatricians as having an inconsequential problem.[35,63] Pediatricians frequently tell children and parents that the chest wall deformity is only cosmetic and will resolve spontaneously. For this reason, the authors have sought to quantify psychosocial functioning with psychometrically sound assessments and to detect the effects of surgical correction of PE by this method. In collaboration with a psychologist with expertise in body image issues, a test for body image effects specific to PE was developed and validated.[34,37] Other researchers subsequently used this or similar psychometric testing tools and corroborated the findings.[15,64] Using the carefully developed and administered instrument, the multicenter study of PE evaluated more than 300 children younger than 21 years before and after operation. Marked improvement in psychosocial functioning was identified postrepair.[37] Several investigators have reported that the severity of the depression as measured by CT scan did not correlate with the patient's or parent's perception of body image concerns, which confirmed previous work on this topic.[65–67]

PREOPERATIVE CONSIDERATIONS

After confirming that the patient's condition is severe enough to fulfill the criteria for surgical correction as outlined earlier, several other factors need to be considered (**Box 1**; **Fig. 2**).

Age

The minimally invasive repair has been performed successfully on patients from age 1 year to older than 50 years.[6,8,10,12,14–19] The ideal age is just before puberty because at that age the chest is still malleable, the support bar is in place during the pubertal growth spurt, the recovery time is short, and the incidence of recurrence is low.[9,20] Patients younger than 8 years also have an excellent result and short recovery time, but because the support bar is removed before the pubertal growth spurt, there is a potential for recurrence.[9,12,38] However, if a young patient has significant cardiac and/or pulmonary compression, an early repair is justified, but the bar should be left in situ for 3 years. 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

Box 1
Indications for operation

An operation is indicated if 2 or more of the following criteria apply:

Chest CT shows cardiac and pulmonary compression or both and a CT index of 3.25 or greater.

Cardiology evaluation demonstrates cardiac compression, displacement, mitral valve prolapse, murmurs, or conduction abnormalities.

Pulmonary function study shows restrictive and/or obstructive lung disease.

Previous repair has failed.

From Kelly RE Jr. Pectus excavatum: historical background, clinical picture, preoperative evaluation and criteria for operation. Semin Pediatr Surg 2008;17(3); with permission.

be inserted by using a chest tube switch technique or later if the condition recurs. Recurrence rate is less than 5% (**Table 2**). During the first decade, the minimally invasive procedure was used only in prepubertal patients, but experience has shown that postpubertal patients tolerate the procedure well; excellent results have been reported in patients in their 30s and 40s.[9,12,15–19] The older patients require 2 or more bars in more than 50% of the cases.[9,12]

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 those with severe deep, asymmetric, Grand Canyon–type depressions are more of a challenge and often require 2 bars. Park and colleagues[6] and other researchers[7] have suggested using an asymmetric bar in these patients. In patients in whom the depression mostly involves the upper chest, care needs to be taken not to place the bar too high because such placement will interfere with the axilla and its vital structures. Patients who have mixed excavatum and carinatum deformities may have residual protrusion of the carinatum after 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.[17,67,68] Patients who have Currarino-Silverman syndrome or 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. Therefore, the minimally invasive procedure is not recommended in this category of patients.

Preoperative Preparation

If the patient has a history of nickel allergy, which occurs in 2% of the population, a titanium bar should be used.[69] The titanium bar needs to be ordered from the manufacturer before surgery (Biomet Microfixation, Jacksonville, FL, USA).

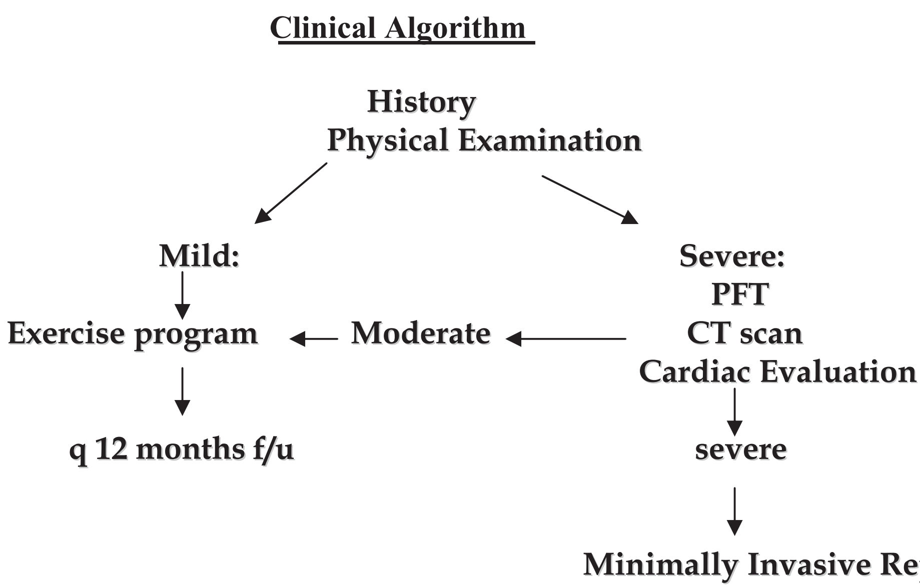

Fig. 2. Indications for PE repair. f/u, follow-up; PFT, pulmonary function test. (*From* Kelly RE, Cash TF, Shamberger RC, et al. Surgical repair of pectus excavatum markedly improves body image and perceived ability for physical activity: multicenters study. Pediatrics 2008;122:1218–22; with permission.)

Table 2
Long-term results after bar removal

Overall Results for Primary Repairs	
Total number of primary repairs	1235
Total number with bar removed	903
Excellent result	773 (85.6%)
Good result	99 (11.0%)
Fair result	11 (1.2%)
Poor result	6 (0.7%)
Failed	11 (1.2%)
Bar removed elsewhere	3 (0.3%)

Data collected through December 18, 2009.

The manufacturer needs to know the length of the bar required, and a copy of the CT scan at the insertion site is required so that the bar can be prebent.

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 preoperatively.

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. An alternative method includes elevating the torso on a mattress and extending the arms posteriorly.[22,70] This position allows insertion of the thoracoscope superior to the incision site but has the disadvantage of overextending the chest during the surgery. Another alternative position is to flex the left shoulder and elbow anteriorly, adjacent to the head,[71] 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.[11,12,23,71–73] Most surgeons use right-sided thoracoscopy,[12,23] others prefer left-sided thoracoscopy,[71] some use bilateral thoracoscopy,[74,75] and some insert the scope and introducer through the same thoracostomy sites.[76] In patients with extremely deep depressions, it may be necessary to use bilateral thoracoscopy because the heart is not only compressed but 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[13,71] or even superior to the incision site when the arm is extended posteriorly.[22,40] 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. The authors use blunt instrumentation for trocar insertion and direct the trocar in a superior direction to avoid the liver and diaphragm.[23]

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

The carbon dioxide (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 2 bars are being inserted, there is 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. It was therefore decided to insert the bar through 2 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 (Langer 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 2 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 positions and extended laterally if necessary. The inframammary incisions give excellent access to the anterior chest wall, even allowing insertion of 2 bars, and give an excellent cosmetic result because 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, the patient requires 2 bars: one under the sternum and the other 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 to keep the introducer at such a position, 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[77] or by lifting it with a towel clamp or heavy suture.

An extrapleural approach has been advocated by Schaarschmidt and colleagues[74] to prevent pleural and pericardial reaction, with good preliminary results. This approach 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 PE. Repeating this lifting maneuver numerous times loosens up the anterior chest wall, prevents substernal trauma, intercostal muscle injury caused by bar rotation, and minimizes the pressure on the bar, which decreases the risk of bar displacement and postoperative discomfort. The PE 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 footplate was developed and attached to the bar to give it more stability.[11] Initially, the stabilizer was held in position only with fascial sutures, but it frequently became detached from the bar, so a wire suture was used to lash the stabilizer to the bar. Recently, the wire has been replaced with a braided polyblend suture called the FiberWire (Arthrex Inc, Naples, FL, USA), which is soft and much easier to apply than the wire. However, even with a stabilizer attached, the bar is dislodged in some patients during the first 3 weeks before scar tissue can be laid down, and therefore additional support is needed during those first few weeks after surgery. Hebra and colleagues[10] were the first to advocate placing a suture around the bar and underlying ribs and called it the "third point fixation." The investigators 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.[8,9,78] Some centers use wire instead of absorbable sutures,[6,25,69,74] which increases the risk of injury to the underlying lung, especially if the wire fractures.[68]

NUMBER OF BARS

Initially, the procedure was done only on young patients, so only 1 bar was necessary.[1] However, now that the procedure is being used more commonly in postpubertal patients, numerous investigators have reported that 2 bars give better and more stable results.[1,8,9,12] Patients with Marfan syndrome, asymmetric Grand Canyon–type deformities, and wide saucer-shaped deformities; older or postpubertal patients; and patients in whom the procedure needs to be repeated also usually require 2 bars.[1,7,9,12,67] A second bar should be inserted if the repair is suboptimal after insertion of 1 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. The authors have never regretted placing a second bar but have often regretted placing only one.

BAR AND CHEST CONFIGURATION

Many patients require a reoperation because the condition was 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 be semicircular 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 bent only at each end (table top configuration) gives 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.[7,26] In patients with asymmetric deformities, Park and colleagues[6] have recommended using an asymmetric bar, which gives more lift on the side of the asymmetric deformity.[8]

REOPERATION

Reoperations on failed previous repairs have been successfully accomplished in 51 previous Nuss repairs, 39 previous Ravitch repairs, 4 failed Ravitch and Nuss repairs, and 3 previous Leonard repairs. Thoracoscopy is particularly important in these groups of patients because they usually require lysis of adhesions before the tunneling can commence, which requires additional port placement.[75,79] A postoperative chest tube is helpful in managing the inevitable lung leak and oozing that follows the lysis of pulmonary adhesions. The failed Ravitch procedures may be classified into 2 categories, those in which there was a simple cave-in after the open repair, which is easily corrected with a substernal bar, and those in which there is diffuse osteochondrodystrophy, a condition that is not amenable to correction with a substernal bar because of excessive calcification and rigidity of the chest wall. In this group of patients in whom the Ravitch procedure failed and there was severe recurrence, acquired asphyxiating chondrodystrophy, and a rigid chest wall, there were 2 cases of temporary arrhythmic arrests that were thought to be caused by pressure of the introducer on the heart. Both patients were resuscitated, but in one patient sternotomy was required because the rigid calcified chest wall did not allow external cardiac compression. Complications are higher in the group of patients requiring reoperation: 55% required chest tubes, 8% had hemothorax, 8% had pleural effusion requiring drainage, and 2 had temporary arrhythmic arrest as mentioned earlier. The results were excellent in 66%, good in 30%, and fair in 2%, and the procedure failed in 2%, which is less satisfactory than the primary repair group.[3,67,79]

Prior thoracic surgery and concomitant open or thoracoscopic intrathoracic procedures have been successfully performed in conjunction with the minimally invasive PE repair.[80]

PAIN MANAGEMENT

A preemptive pain management protocol is used to prevent the pain cascade from being triggered. All patients 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, midazolam is administered orally 45 minutes before the patient is taken to the operating room. After induction of anesthesia, a dose of intravenous ketorolac is given, and this drug is continued every 6 hours until day 3. While the patient is receiving the ketorolac, the intravenous 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 postoperative days 1 and 3. Until recently, an epidural infusion of fentanyl and bupivacaine was used, which was started during induction of anesthesia and continued until postoperative day 3 or 4. However, because 2 of 1300 patients developed partial paralysis, patient-controlled analgesia pumps are now used postoperatively. Patients also receive low-dose diazepam for muscle relaxation and anxiolysis. Stool softeners and laxatives are used prophylactically starting on day 1 to prevent constipation. Patients are discharged on day 4 or 5, with oxycodone and ibuprofen for pain control and diazepam and Robaxin for muscle relaxation.

Patients may return to school whenever they are strong enough; the duration varies with age, because prepubertal children recover quicker and are usually ready to return to school in 2 weeks, whereas postpubertal patients require 3 weeks. All patients are restricted from participating in sporting activities for 6 weeks, at which time they may recommence aerobic activities, and competitive sports may be resumed at 12 weeks postrepair.

EARLY POSTOPERATIVE COMPLICATIONS

Early complications (**Table 3**) 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 the complication rate after the early learning experience.[11,13,20,21,24]

An insignificant apical pneumothorax secondary to CO_2 insufflation for thoracoscopy is usually present on the initial chest radiograph and resolves spontaneously. A chest tube was inserted in 3% of the 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. These pneumothoraxes resolve spontaneously and are really a part of the operation rather than true complications because there is no lung leak in a primary pectus repair. However, redo operations with lysis of pulmonary adhesions frequently do require a postoperative chest tube

Table 3
Early postoperative complications of patients undergoing primary surgery

Complication	% (Number of Patients)
Pneumothorax with spontaneous resolution	66.0 (746)
Pneumothorax with chest tube	4.4 (50)
Horner syndrome with spontaneous resolution	15.2 (188)
Drug reaction	3.2 (39)
Suture site infection	0.9 (11)
Pneumonia	0.7 (8)
Hemothorax	0.5 (6)
Pericarditis	0.5 (5)
Pleural effusion (requiring drainage)	0.3 (4)
Temporary paralysis	0.1 (1)
Cardiac perforation	—
Death	—

Data collected through December 18, 2009.

because these patients do have an air leak.[75,79] Pneumonia is rare in these young patients (0.5%), but postoperative incentive spirometry is vigorously encouraged, and all patients are given prophylactic antibiotics (cefazolin) for 24 hours. Wound and/or bar infection can be prevented if all the precautions for foreign body insertion are meticulously adhered to, and such infections have occurred in less than 1% of patients.[81–83] 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 erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) level have returned to normal.[81–83] There have been reports of an increased infection rate on the side with the stabilizer,[84] but that has not been encountered by the authors in more than a thousand cases.

Pericarditis has occurred in 0.5% of the patients.[69,85,86] The cause is unclear. The condition appears to be caused by nickel allergy because it has not been seen in more than 6 years when screening patients more closely for nickel allergy was started. These patients present with persistent central chest pain, malaise, lethargy, and a pericardial friction rub. If an echocardiogram confirms the presence of pericardial fluid, the patient should be treated with a short course of prednisone. A pleural effusion that lasts for more than 4 days may also be caused by nickel allergy and should be treated similarly after aspirating fluid for culture. If symptoms recur after prednisone has been discontinued, the patient should be tested for nickel allergy (T.R.U.E Test by Allerderm Laboratories, Inc, Phoenix, AZ, USA). If the result is positive, low-dose prednisone can be given on alternate days until the ESR and CRP return to normal levels; if this option is unsuccessful, the steel bar is replaced with a titanium bar, which has only been necessary in 2 patients out of 1123 repairs.

Cardiac perforation has occurred in several centers during the early learning experience and before thoracoscopy was widely available.[6,87–90] Reviewing the preoperative CT scan to determine the position of the heart and its relationship to the sternum is helpful in planning the procedure, especially in patients with severe asymmetry and sternal torsion. If it appears that the heart is severely compressed, elevating the sternum with a hook or suction cup during the tunneling greatly minimizes the risk of injury. In addition, first tunneling 1 or 2 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 anteroposterior 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.[88]

LATE COMPLICATIONS

Bar displacement has been the biggest late challenge. The initial bar displacement rate was 18.5% (**Table 4**). After the introduction of stabilizers, the rate dropped to 7.4%, and with the addition of pericostal sutures placed around the bar and underlying ribs, the rate dropped to 2%, only half of whom required revision (**Fig. 3**).[9,12,20]

The standard procedure is to place a stabilizer attached to the bar, with FiberWire suture on the left and multiple double-stranded “0” polydioxanone pericostal (PDS) sutures around the bar and underlying ribs on the right. If feasible, pericostal sutures are also placed on the left. If displacement is less than 20°, it can be observed by obtaining

Table 4
Late postoperative complications of patients who had primary surgery

Complication	Number of Patients	Percentage
Bar displacements	70	5.7
Requiring revision	50	4.1
Overcorrection	42	3.4
Bar allergy	38	3.1
Wound infection	18	15
Recurrence	13	1.1
Hemothorax	2	0.2
Skin erosion	1	0.1
Accidental death[a]	1	0.1

Data collected through December 18, 2009.
Total number of patients, 1235.
[a] Accidental death occurred after 3.5 years.

another chest radiograph in 1 month, and if there is no further progression, surgical revision is not usually required; however, if the displacement is more than 20° or it occurs right after surgery, it needs correction. At present, less than 1% of the patients require revision because of bar displacement.

Nickel allergy, which is present in 2%[69,85] of the population, may manifest early with pericarditis or persistent pleural effusion or may 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 give a negative result and testing for nickel allergy gives a positive result. Treatment consists of local wound care and a short trial of prednisone. If the patient responds, administration of low-dose alternate-day prednisone until the ESR and CRP are back to normal levels usually resolves the problem. If the patient responds to the steroid therapy, the bar is left in place until it is time for removal. If the patient does not respond to treatment, the steel bar needs to be replaced with a titanium bar.[69,85] In the series of 1123 patients, 35 patients (3.1%) had allergy, of whom 22 were diagnosed preoperatively and received a titanium bar. Of the 13 who had a steel bar inserted, 10 were treated successfully with prednisone and 3 required bar removal.

Overcorrection, resulting in pectus carinatum, has occurred in 0.4% of the patients. These patients had Marfan syndrome and very deep cup-shaped deformities. Early bar removal was successful in 1 patient, and 2 required an external pressure brace. Other researchers have reported on carinatum developing especially in patients with asymmetry and a twisted sternum.[25]

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 caused by bar displacement, stabilizer dislocation, bar being too tight or too long, sternal or rib erosion, infection, or allergy. An anterior and lateral chest radiograph, complete blood cell count, ESR, CRP level, and T.R.U.E. Test for allergy will identify the cause and allow appropriate treatment.

One accidental death occurred 3.5 years after pectus repair and was unrelated to the pectus surgery because the patient fell off an eighth-story balcony during a graduation celebration.

RESULTS

Long-term results 1 year after bar removal in the 903 patients who underwent primary repair are excellent in 773 patients (85.6%), good in 99 (11.0%), fair in 11 (1.2%), and poor in 6 (0.7%); the procedure failed in 11 patients (1.2%) and bar removal was done elsewhere in 3 patients

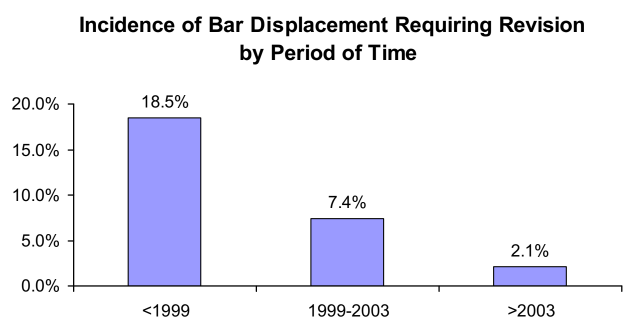

Fig. 3. Improvement in bar displacement rate over time as new modifications were introduced.

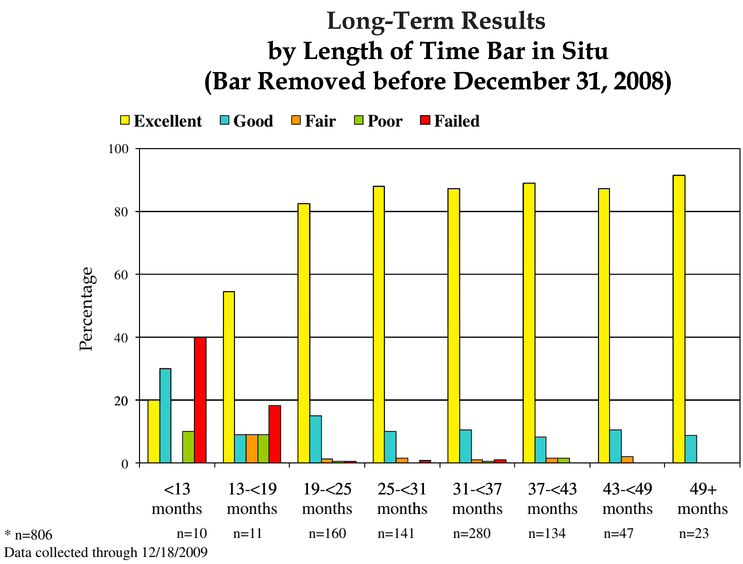

Fig. 4. Long-term results show that the bar should remain in situ for 2 to 4 years. The longer the bar remains in place the lower the recurrence rate up to 4 years.

(0.3%). (see **Table 2**; **Figs. 4** and **5**). Similar results have been reported by other centers.[5–8]

If the bar is removed before 2 years, the recurrence rate increases inversely with the length of time the bar remains in situ (see **Fig. 4**).

The age at the time of repair affects the recurrence rate. If the repair is performed in the very young, aged 5 years or younger, there is an increased risk of recurrence, although age is not as important as the duration of bar placement and should not prevent repair in a young patient with a severe PE with cardiac or pulmonary compression (see **Figs. 4** and **5**).[9] The higher recurrence rate in children aged 5 years and younger is mostly because these patients underwent repair during the learning curve, when patients did not keep their bars in place for the now standard 2 to 4 years.

All patients are encouraged to exercise regularly starting 6 to 8 weeks postoperatively. It is thought 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.

Postoperative cardiopulmonary function has been shown to have good improvement in some

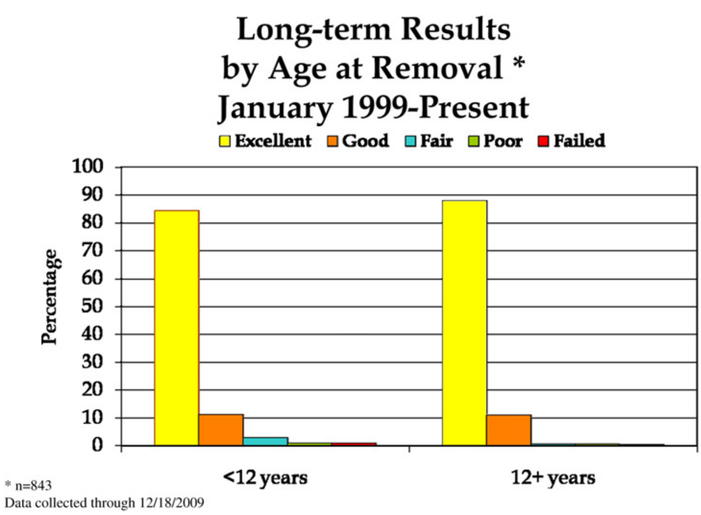

Fig. 5. Graph showing slightly improved results in older patients.

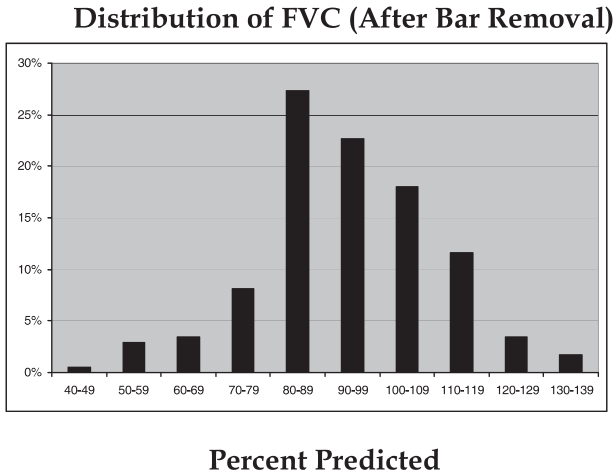

Fig. 6. Postoperative resting pulmonary function studies showing that the peak for FVC has shifted toward normal. (*From* Nuss D. Minimally invasive surgical repair of pectus excavatum. Semin Pediatr Surg 2008;17 (3):215; with permission.)

studies and less so in others. Several studies have shown significant improvement in pulmonary function postoperatively,[27–29,31–33,40] whereas others have shown no significant change.[19] 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 PE, and whether the studies were done during exercise or at rest. In the authors' series of more than 900 cases, the preoperative resting pulmonary studies showed a marked shift to the left and a significant correction postoperatively (see **Fig. 1**; **Fig. 6**)[26,31]:

Postoperative cardiac studies have shown an increase in cardiac filling and stroke volume.[30–33] There is a discrepancy between the overwhelming number of patients reporting dramatic improvement in their exercise tolerance and the results of cardiac and pulmonary studies. Surgeons regularly hear "I never realized how incapacitated I was until I had my pectus corrected," "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 after the minimally invasive repair.[8,15,34–36]

BAR REMOVAL

The bars 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 with the bar in place (see **Fig. 4**). There have been a few patients who have kept their bar in situ for 4 or more years without any problems. If patients grow more than 6 in (15 cm) after bar insertion and become symptomatic with lateral chest pain, they need to be evaluated to see if early bar removal is required.

Bar removal is accomplished under general anesthesia with positive pressure ventilation and 5 to 6 cm of positive end-expiratory pressure to prevent pneumothorax. Both sides of the bar should be mobilized, and the bar should be unbent using either the bar flippers or Multibenders (Biomet Microfixation, Jacksonville, FL, USA).[75,91,92] After straightening, the bar is removed slowly while monitoring the ECG and all the other vital signs. A postoperative chest radiograph is obtained routinely to check for pneumothorax. The complication rate for bar removal in 815 patients was 3 pneumothoraxes requiring aspiration and 1 wound infection. Elsewhere, there have been isolated reports of major complications, including 1 cardiac arrest and a pulmonary hemorrhage requiring thoracotomy.[90,93]

SUMMARY

In the 22 years since the first minimally invasive PE 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 a 95% overall patient satisfaction rate.[12,15]

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