Valuable lessons from two decades of pectus repair with the Willital–Hegemann procedure

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Objective: Pectus deformities are the most common congenital hereditary chest wall deformity. The aim of this study was to evaluate the efficacy of thoracic wall reconstruction using a uniform technique of internal stabilization with stainless-steel struts.

Methods: Hospital charts of patients with chest wall deformities managed with the Willital–Hegemann procedure between January 1984 and January 2004 were reviewed.

Results: Surgical corrections were performed in 1262 patients with pectus deformities (968 male and 294 female patients). The corrections were completed with successful repair in 1244 (98.6%) patients, along with a low complication rate of 5.7%. The median age of the patients was 14.9 years (range, 2–53 years). The follow-up period ranged from 2 to 12 years (mean, 5.4 years). Major recurrences were observed in 18 (1.4%) patients, and mild recurrences were observed in 46 (3.6%) patients. There was 1 death in this series. The struts were removed after a period of 24 to 36 months and were associated with a complication rate of 2.6% at the time of removal.

Conclusion: Custom-tailored molding of the chest wall can be achieved by using this method, which is not possible with minimal-access techniques. Open repair is effective for all variations of chest wall deformities and in patients of all ages, causes only mild pain, and produces good physiologic and cosmetic results. Improvement of subjective complaints, satisfactory long-term results, and improvement in psychological problems indicate the need to offer this procedure among other surgical correction options for low-risk children.

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Received for publication Jan 22, 2007; revisions received June 12, 2007; accepted for publication June 15, 2007.

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J Thorac Cardiovasc Surg 2007;134:871-6
0022-5223/\$32.00

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doi:10.1016/j.jtcvs.2007.06.008

Pectus excavatum (PE) and pectus carinatum (PC) deformities are the most common major congenital anomalies, with an incidence of 1:400 in white male births.¹ An increasing number of reports from previous years indicate that both PE and PC deformities cause physiologic limitations and have adverse cosmetic and psychologic effects.²⁻⁴ The patients with both deformities compensate for diminished chest wall excursions during respiration through the use of wider diaphragmatic excursions and tachypnea. However, patients with severe PE associated with cardiac displacement frequently experience tachycardia to compensate for reduced cardiac output. Chest wall depression is well tolerated in infancy; however, many older children report subjective complaints, such as dyspnea, cardiac dyesthesia, and limited work performance. In the majority of patients, these symptoms seem to deteriorate until a steady state of completed pubertal growth has been achieved. Most pectus deformities maintain the same degree of severity throughout adult life, and, in many cases, will gradually become more symptomatic.⁵

Except for severe chest wall deformities, it is generally extremely difficult to predict the course of progression after the deformities have been discovered in early

Abbreviations and Acronyms

PC = pectus carinatum
PE = pectus excavatum

infancy. Therefore, mild forms of abnormalities warrant the wait-and-watch approach during the first 4 to 5 years of life before operative management can be considered. The management of these deformities, however, has gained more importance in the past decade because of the patients' increased participation in competitive school sports, as well as in athletics, and the changing trends in the world of fashion, which expose more of the chest to the public view. It is important to note that although thoracic wall deformities do not arouse the sympathy generated by limb or cranial anomalies, children with such deformities are viewed with curiosity by their classmates and are often confronted with teasing remarks. Surgical correction has been performed in many centers, mainly for aesthetic and psychological reasons; however, compression and secondary changes of the intrathoracic organs have been indications for surgical intervention.

Until the end of the last decade, only a few centers performed corrective operations for pectus deformities, with a variety of techniques based on the early experiences of Ravitch⁶ and Welch.⁷ The reported results have been inconsistent, which has often caused reluctance among referring physicians, as well as in patients, to pursue correction of pectus deformities. The report by Hegemann and Leutschacht⁸ on surgical management with metal struts to stabilize the sternum in 1965 was modified by Willital⁹ in 1981 and laid the basis for the operative techniques evaluated in this series. Since 1998, with the new concept of minimally invasive PE repair reported by Nuss and colleagues¹⁰ and the availability of several informative pectus Web sites that advertise this procedure, many patients of all ages have become aware that their deformities can be corrected with a higher degree of success and lower risk of morbidity than has occurred in previous years. The present report summarizes the clinical experience with the Willital–Hegemann technique and discusses the modification made in open repair for both PE and PC deformities in 1262 consecutive patients during the past 20 years. Consent was obtained from patients for data evaluation.

Materials and Methods

At the Pediatric Surgical University Clinic, Münster, Germany, 1262 surgical corrections were performed on patients with congenital chest wall deformities from January 1984 through January 2004. During this time, 2780 patients were referred to the center for the evaluation of chest wall deformities, and a thoracic reconstruction operation was performed on 1031 (81.7%) patients with PE and 138 (10.9%) patients with PC. Other forms of pectus

deformities were documented, and operations were performed in 93 patients. Pectus deformities occurred more frequently in male (968 patients) than female (294 patients) patients. The records demonstrated that in 81% of the patients, the pectus defect was evident within the first year of life.

Classification

The patients were graded according to the Willital classification, which is based on morphologic findings of the thorax and divides congenital chest wall deformities into 11 types: funnel chest (4 types), pigeon chest (4 types), combination of funnel and pigeon chest, chest wall aplasia, and cleft sternum.¹¹ This classification allows assessment of the operative technique with regard to the location of implantation of the metal struts, as well as the determination of the number of struts to be used for internal stabilization of the chest wall. It also takes into consideration the asymmetric presentation of pectus deformities, along with inclusion of the rare forms. Evaluation of 1262 patients with pectus deformities demonstrated that 67.7% of the patients have PE deformity with a symmetric chest wall (Figure 1).

Surgical Technique

The standard surgical technique was the Willital–Hegemann procedure, which was described in our previous report on PE correction.¹² Briefly, this technique uses stainless-steel struts for the correction of the deformity. A vertical midline incision was preferred in male patients. In female patients a submammary incision that is curved upward at the midpoint is preferred, thus avoiding the complication of breast deformity and impaired breast development.¹³ In the incision skin, fat, and pectoral muscles were reflected in a single flap, and the entire dissection was performed with needle-tipped electrocautery. The pectoral muscles were severed from its insertion at the edge of the sternum and costal cartilage to expose the entire impression of the deformity, generally formed by 5 to 8 pairs of ribs (third to tenth rib). When the deformed costal cartilage had been completely exposed, the perichondrium was incised with the needle-tip cautery in the form of an H (Figure 2, A). The subperichondrial dissection was carried out with small, sharp periosteal elevators, and the perichondrium was scraped away from the underlying cartilage. The deformed costal cartilages were resected parasternally from its junction with the rib to within 1 cm of the sternum, as well as at the level of transition to the normal ribs, leaving the uppermost normal cartilage intact. The attachment of the rectus muscle to the sternum was severed, and the sternum was clamped with a Kocher clamp and dissected free from the anterior mediastinal tissue after retrosternal mobilization. A partial transverse sternal wedge osteotomy was performed, if required, at the angle of Ludovici. Once the sternum was dissected free, as described, a perforated Hegemann steel strut (Lettenbauer, Erlangen, Germany) was passed transsternally, with its edges resting anteriorly on the ribs (Figure 2, B). The strut was bent in such a way that it fit the thorax wall perfectly at the edge of the impression. Two parasternal metal struts were also used, with the points of fixation being the second rib and the lowest end of the rib cage (Figure 3). The transsternal strut was fixed to the 2 parasternal struts with stainless-steel wires for additional support. The 2 parasternal struts also provided anchorage to the completely mobile chest segments, which were formed as a result of double-

CLASSIFICATION – CHEST WALL DEFORMITIES

Figure 1. Percentile distribution of 1262 patients undergoing operations (1984–2004) and schematic representation of chest wall deformities according to the Willital classification. *PE*, Pectus excavatum; *PC*, pectus carinatum.

bilateral chondrotomy. Heavy resorbable suture material was then used to close the sternal osteotomy, as well as to suture the ends of the costal cartilages. Two single-limb chest tubes were placed in parasternal positions at the level of the highest costal cartilage resection. The pectoral muscle flaps and the severed rectus muscles were then sutured and fixed to the sternum. The overlying subcutaneous and cutaneous structures were finally united in the conventional manner to restore normal chest wall anatomy.

Perioperative antibiotic therapy was administered, with ceftriaxone being the drug of choice. All patients were also administered strong analgesics for the first 24 to 72 hours. The chest tubes were generally removed on the third day or when the drainage was less than 25 mL for a 12-hour shift. Wound infections were rare, and the patients are mobilized after chest tube removal. On the fourth day, pulmonary exercises are commenced. At the time of discharge, patients were advised to avoid body-contact sports until the struts had been removed. Regular swimming was encouraged, along with light athletic exercises. The patients were followed up at regular intervals (3, 6, 12, and 15 months) after the procedure. The struts were removed after a period of 12 months in patients older than 12 years but were retained for 15 to 24 months in younger children, so as to stabilize the thorax

that was still undergoing a growth spurt. After strut removal, the patients were followed up to 3.5 years, and if necessary, longer follow-up periods were carried out.

Results

Surgical corrections were performed in 1262 patients with pectus deformities (968 male and 294 female patients). The median age of the patients was 14.9 years (range, 2–53 years). The corrections were completed with successful repair in 1244 (98.6%) patients, with satisfactory long-term results. The postoperative course of 1261 patients was generally uneventful, and the repairs were completed with a low complication rate of 5.7% (Table 1).

Three struts were used as part of the standard procedure; however, in 35 (28%) patients with severe deformities, additional struts (transsternal) were required. Blood transfusions were required in 4 patients. The mean hospital stay from 1984–1999 was 20.5 days (including 2 weeks of physiotherapy exercises), which was reduced to 6.2 days

Figure 2. A, Multiple reactions of deformed lower costal cartilage and ribs at the culmination point. B, Stabilization of the mobile sternum with a transsternal metal strut with sutures and anastomoses of the ribs with heavy resorbable sutures. *Inset*, Transsternal strut.

from 1999–2004. The struts were removed after a period of 24 to 36 months and were associated with a complication rate of 2.6% (wound seroma) at the time of removal.

The follow-up period ranged from 2 to 12 years (mean, 5.4 years), and the documentation of the preoperative deformity, as well as the status of surgical repair, included (1) measurement of thorax diameter with a pelvimeter, (2) electrocardiographic analysis, (3) pulmonary function tests in children older than 8 years, (4) chest ultrasonographic analysis, and (5) clinical

photography. Since 1994, the documentation included video stereoraster examinations. Major recurrences were observed in 18 (1.4%) patients and required operations. Mild recurrences were observed in 46 (3.6%) patients. There was 1 death caused by intraoperative cardiac arrest in a patient with a previously known cardiac anomaly. In 1223 (97%) patients records at the time of follow-up examinations documented that the subjective complaints of the patients before surgical intervention were eliminated.

Figure 3. Schematic overview of the operative technique showing strut placement.

Table 1. Surgical complications after pectus repair: 1984–2004

Complications	No. of patients
Serothorax	29
Pneumothorax	16
Wound dehiscence	8
Wound infection	7
Strut dislocation	7
Hemothorax	2
Wound hematoma	1
Osteomyelitis	1
Intraoperative cardiac arrest	1

Discussion

The use of minimal-access surgery in pectus repair has gained popularity, and the indication for PE has increased in centers worldwide.¹⁴ Minimal-access surgery is well suited for patients with type 1 PE; however, the repair of deep type 1 PE deformity can be technically difficult with this approach.^{15,16} However, in asymmetric forms of type 2 and type 4 PE, the results of minimal-access pectus repair are variable and not always satisfactory. Also, if the depression is around the area of the xiphoid, it is quite difficult to place the bar under the deepest point of the pectus. Furthermore, in combined forms of pectus deformity type 9 (PC combined with PE), lifting the pectus deformity in the minimal-access procedure accentuates the associated PC deformity even further, with unsatisfactory results. Furthermore, the increased rate of infection associated with minimal-access procedures in most of the series reported was not observed in our open repair series.^{17–19} Minimal-access surgery is limited in its approach to PC.

There is no unanimity in the literature as to the best time of intervention, as well as the necessity of such an intervention.²⁰ Surgical intervention of pectus deformities is easier to perform in children because the ribs are soft and easier to correct; however, repair of both PE and PC in preadolescent children is recommended for only those few who are symptomatic because the amount of deformed cartilage commonly increases during the pubescent period of skeletal growth, as does the severity of the deformity. Furthermore, in female patients it is advisable to wait until the pubertal breast growth demarcates the submammary line to avoid breast deformities caused by incision scars. In this series of patients operated on at less than 12 years of age, there was no increased correlation to recurrence or recurrent deformation caused by the growth spurt later in the adolescent stages.

The preservation of the perichondrial sheaths during resection of the deformed cartilage must be stressed.²¹ The absence of perichondrial sheath caused by resection or damage can lead to the regeneration of a malformed rib. In

our patients we encountered no problems after perichondrial damage in adolescents, and the results were good; however, in children younger than 12 years, this could be the reason for mild recurrence. Furthermore, damage to the perichondrium produces calcified, rigid, and inflexible bone-like tissue, resulting in the chest wall becoming a near-rigid object, with respiratory movements largely dependent on diaphragmatic excursions. Not only is the preservation of the perichondrium important, but it is equally important to suture the perichondrial sheaths to prevent the regenerating cartilage from becoming a fused bony plate instead of individual ribs, as observed in many patients during the time of strut removal.²² Damage to the perichondrial sheaths could explain the significant reduction in the total capacity and inspiratory vital capacity of the lungs that could result from decreased compliance of the fused chest wall after pectus surgery.^{23,24}

Because of the large number of cases referred for surgical intervention, it was important to rule out allergy to metal. In all patients a careful history was taken, and in suspected but unconfirmed cases, a skin test was performed as part of the preoperative workup. In one patient with nickel allergy, titanium struts were used, along with non-resorbable sutures.²⁵ In the initial part of the series, the metal struts were placed on the sternum and were held to the sternum by using multiple metal wires. These wires posed technical challenges during strut removal. This practice was given up, and metal struts were introduced through the sternum (transsternal). Also, only 2 metal wires were used to secure the transsternal to the parasternal struts. This standardized the placement of the strut for PE, as well as PC, repair. To place the transsternal strut, it was mandatory to mobilize the sternum from its mediastinal attachments, which is presumed to be an important factor in the prevention of recurrences.²⁶ Routine use of the sternal strut for patients with both PE and PC minimizes movement of the anterior chest wall with respirations and physical activities during the postoperative period, which reduces pain, thus decreasing the need for analgesic medications. The strut also stabilizes the chest, which decreases the possibility of a recurrence and is predictive of a long-term good result.²⁷ In patients with asymmetric pectus deformities and those with a large chest area, additional struts have been used to provide stability to the flail chest wall segments.²⁸

In severe pectus deformities, not only the cartilage but also the ribs are deformed. The deformities of the ribs are more prominent in PC than in PE, and the affected ribs are generally in the middle or lower thorax. To get excellent surgical results, along with bilateral parasternal chondrotomy, rib osteotomies were performed, when necessary, to remodel the rib cage. A partial rib osteotomy was performed with bone pliers, and the weakened rib was fractured under digital pressure. The fractured ribs were then stabilized with

resorbable figure-of-8 sutures. Our experience has shown that the osteotomy of the ribs is not associated with increased pain or discomfort in patients postoperatively if secured by figure-of-8 sutures. However, in the absence of figure-of-8 sutures, pain was experienced by the patients when they took a deep breath because of friction of the rib fragments. Also, friction of the fragments generates increased bone formation, which is palpable at the point of fracture. Surgical intervention was not required in any of our patients to treat this problem.

Earlier in our series, chest tubes placed during surgical intervention were guided out through the umbilicus for aesthetic reasons. Inadequate draining of the intrathoracic space caused by loss of length of the chest tubes during supraumbilical transit was responsible for the larger number of intrathoracic air and fluid accumulations in our series. Postoperative abdominal distention caused by peritoneal irritation was frequently observed as a result of supraumbilical chest tube transit. Since 1994, the chest tubes have been guided out through the intercostal space, and reduction of these complications has been observed.

The main reasons for recurrence in this series were (1) inadequate mobilization of the depressed chest wall, (2) unstable fixation of the mobilized chest wall, (3) no or inadequate remodeling of the lowest parts of the deformed ribs, (4) no axial anastomoses of the ribs, (5) postoperative infections, and (6) postoperative trauma. Complete exposure of the sternum and ribs is necessary for satisfactory results, because it allows complete intraoperative assessment of the deformity and is of significant importance in patients older than 15 years who might require an additional transsternal strut because of the extensive area of sternal depression.²⁹ This also allows placement of struts and stainless-steel wires under complete vision and avoids serious complications, such as cardiac perforation.³⁰

In conclusion, the experience with the Willital-Hege-mann procedure over 2 decades has yielded excellent long-term results, along with a low rate of recurrence. Furthermore, this procedure is applicable for the management of all types of pectus deformities.

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