Staged management of pectus carinatum

Amy S. Cohee, James R. Lin, Frazier W. Frantz, Robert E. Kelly Jr.

Abstract

Aim: The aim was to report the treatment of pectus carinatum with a novel Argentine brace and operation.

Methods: The bracing and clinical data of 137 consenting pectus carinatum patients treated between October 2008 and December 2011 were reviewed for outcome. Institutional approval was obtained. Data are reported as median (range).

Results: Median age 122 bracing patients was 14 (10–28) years with 67 (55%) progressing under active treatment. Five patients (4%) were lost to follow-up, and thirteen (11%) failed treatment. Thirty-seven patients (30%) exhibited flattening of the sternum after 6 (1–24) months without surgery. After flattening, patients then wore the brace for progressively fewer hours each day as a “retainer” for 5 (3–19) months. Five patients (4%) experienced recurrence 5 (3–7) months after brace treatment was discontinued. Complications were limited to transient skin breakdown in nine patients. Three of the 13 Argentine brace failures and 15 other pectus carinatum patients were treated surgically. Thirteen underwent Abramson’s minimally invasive operation and five an open repair, all with good initial correction. For Abramson repairs, seven patients have had bars removed, with results rated as excellent (n=4), good (n=2), and failure (n=1, converted to open with excellent result later). In three patients with stiff chests, costal cartilage was resected thoracoscopically during the Abramson repair with measurably improving compliance.

Conclusion: Staged treatment of pectus carinatum allows most teenagers to be managed non-operatively. For patients who fail bracing or are not amenable to bracing, minimally invasive surgical treatment for pectus carinatum is a viable option.

© 2013 Elsevier Inc. All rights reserved.
stiffness and/or severe asymmetry for example), traditional open repair with sternal osteotomy and costal cartilage resection can be employed.

Large clinical series utilizing compressive orthotic bracing for chondrogladiolar pectus carinatum have been reported by surgeons from around the world [2–8]. Martinez-Ferro et al. have advanced this technology with implementation of a dynamic compression system (DCS) [8]. This system, developed in Buenos Aires, Argentina, uses a custom-made aluminum brace and cushioned compression plate, which allow measurement and adjustment of the pressure applied to the sternum by the brace [8]. Successful utilization of compressive orthotic bracing relies upon patient compliance and requires frequent clinical follow-up. Serial brace adjustments are needed to maintain adequate compressive force as the sternum flattens. The benefits of brace therapy include avoidance of surgical risks, postoperative pain, convalescence, and activity restrictions associated with implant placement. No scarring is produced, and there is significant reduction in treatment costs. Advocates of the non-operative approach recommend it as initial therapy for most patients with pectus carinatum [2–8]. This approach is generally supported by the medical community and by health insurance providers in the USA.

Minimally invasive repair of pectus carinatum as described by Abramson et al. has gained increasing international acceptance since its introduction in 2005 (Fig. 1A and B) [1,9]. This technique is associated with high patient satisfaction and minimal complications. It produces immediate sternal flattening, generally in a single operation [10,11]. Abramson’s operation is also suitable when access to close clinical follow-up is limited, in tertiary settings where patients are treated at a distance far from their homes, and where patient compliance (necessary for successful bracing therapy) is questionable. While activity restriction is necessary due to the presence of the metal implant, the advantages of this procedure over traditional open repair are significant. They include limited chest wall dissection with minimal or no resection of cartilage, a lack of incisions over the anterior chest wall, and shorter operative time.

In this report, we summarize our three-year experience with staged management of pectus carinatum, utilizing bracing therapy as initial treatment for the majority of patients and reserving surgical treatment with the Abramson repair or traditional open repair for patients who fail bracing therapy, those who are unlikely to be compliant with bracing, and those with significant chest wall stiffness and/or severely asymmetric deformities.

1. Materials and methods

A retrospective review was conducted of our prospectively gathered Chest Wall Anomaly Clinical Database. This included all patients presenting to the Children’s Hospital of The King’s Daughters in Norfolk, Virginia, USA for treatment of pectus carinatum between Oct. 2008 and Dec. 2011. Informed consent for the review of clinical data recorded in the database was requested from patients and families upon evaluation. The informed consent process follows the guidelines set forth by the US Department of Health and Human Services, and the Chest Wall Anomaly Clinical Database has the approval of Eastern Virginia Medical School Institutional Review Board (01-05-EX-0175). The use of data for this review was also approved by
the Eastern Virginia Medical School Institutional Review Board (12-01-WC-0006). Quantitative data are reported as median (range), and comparisons were evaluated with the non-parametric Mann–Whitney U test at a significance level of $P = 0.05$.

Between January 2009 and December 2011, 122 patients were braced using the DCS developed by Martinez-Ferro et al. [8]. Chondrogladiolar pectus carinatum was the most common form of pectus carinatum in the cohort, with 117 patients exhibiting this variant. One patient with chondrogladiolar pectus carinatum had undergone surgical repair of pectus excavatum by the Nuss procedure with overcorrection. Five patients had mixed pectus carinatum and excavatum deformities. One of these five patients had a history of a diaphragmatic hernia repair at birth with extracorporeal membrane oxygenation (ECMO) treatment which was followed by the development of a pectus carinatum with a minimal pectus excavatum component.

The treatment algorithm used is depicted in Fig. 2. After initial examination and classification of the pectus carinatum deformity, the compression pressure required to flatten the sternum, called the pressure of correction (POC), was measured in pounds per square inch (PSI). Patients with high correction pressures (POC $> 7.5$ PSI), severe asymmetry, history of previous bracing failure, and those deemed unlikely to be compliant with bracing were offered surgical treatment with an open repair or Abramson repair (including thorascopic rib resection, where appropriate). Patients amenable to bracing were measured for an Argentine brace and fitted at a subsequent clinic. At the time of brace application, the brace configuration was adjusted such that the force on the compression plate, the pressure of treatment (POT), measured $< 2.5$ PSI to minimize the risk of skin breakdown [8]. Physical therapists instructed patients regarding appropriate strength and flexibility exercises to optimize results. Exercises were performed at home according to instructions given by the physical therapist during the
office visit. Each patient was given printed illustrations of the prescribed exercises.

Patients in active treatment were encouraged to wear the brace for as much time as possible, including during sleep but not during athletic activities or showering. A minimum daily bracing duration of 8–12 h was recommended. Daily exercises were strongly encouraged at each visit. Patients in active treatment were seen in the clinic every 1–3 months by a surgeon and physical therapist to make adjustments to the brace and reinforce brace compliance and physical activity. Usually, brace adjustment was done by the physical therapist. After flattening, patients wore the brace for progressively fewer hours as a “retainer” for 5 (3–19) months. Patients in retainer mode were seen approximately every 3–6 months to assess the durability of the flattening, make adjustments to the brace, reinforce brace compliance and physical activity, and consider the possibility of discontinuation of the brace or the need for operative intervention.

The cohort of pectus carinatum patients who underwent operation between Oct. 2008 and Dec. 2011 included three of the DCS bracing-failure patients and 15 other pectus carinatum patients.

2. Results

122 bracing patients [median age 14 (10–28) years] accumulated over three years in our clinic and of these, 67 (55%) were progressing under active treatment. Thirty-seven patients (30%) exhibited flattening of the sternum after 6 (1–24) months without the need for surgery. Five patients (4%) were lost to follow up, and 13 patients (11%) failed treatment.

Five patients (4%) experienced a recurrence at 5 (3–7) months after brace treatment was discontinued. Four of the five returned to wearing the brace as a retainer and continue in active treatment. The remaining patient wore the brace as a retainer for three additional months and experienced durable flattening of the sternum.

No patient had overcorrection to pectus excavatum, though the patient with a mixed deformity and history of diaphragmatic hernia repair with ECMO experienced a deepening of the pectus excavatum portion of the deformity. This patient later underwent a Nuss repair to address the pectus excavatum component, which also corrected the associated marked sternal torsion. All patients who underwent bracing therapy, including those who eventually failed, showed increasing malleability of the chest wall, as measured by declining POC.

Complications of the Argentine brace were limited to transient skin breakdown in nine patients. This was treated with temporary loosening of the brace to lower the POT or discontinuation of bracing until the skin healed completely. Tachycardia associated with cardiac compression prevented bracing in one patient. Computerized tomography showed that the heart occupied the space between the sternum and spine, and thus, the heart would have been compressed by the brace (Fig. 3).

The median initial POC was 4.7 (0.8–10) PSI, indicating moderate flexibility. The median initial POC for patients who failed was 5.5 (2.0–9.5) PSI, whereas the median initial POC for patients with successful correction was 4.7 (2.0–9.5) PSI. The distribution in the two groups differed significantly (Mann Whitney $U = 343, \, n_1 = 37, \, n_2 = 13, \, P < 0.05$ two-tailed). It is noteworthy that we have had a single case with initial correction pressure of >9 PSI who has had an excellent result with brace treatment after 10 months of active treatment and 6 months of retainer mode.

The required time to sternal flattening in active bracing treatment correlated with the initial POC, with patients with higher POCs requiring longer time to flatten. For patients with POC <3 PSI, the time to flattening was 4 (3–7) months. For POC of 3–5 PSI, the time to flattening was 7 (1–24) months. For POC of >5 PSI, the time to flattening was 6 (4–13) months.

Patient-reported daily bracing durations recorded as a measure of treatment compliance revealed that the average daily bracing duration in patients who experienced sternal flattening ($n=37$) was 16 (0–24) h. For patients who wore their braces and failed bracing therapy ($n=8$), the average daily bracing duration was 14 (5–23) h. Four patients in the bracing failure group discontinued use of the brace in the first month after application for various reasons, including pain associated with wearing the brace, skin breakdown beneath the compression plate, inability to sleep in the brace, and lifestyle incompatibility (i.e. self-consciousness in wearing the brace to school).
Three of the 13 Argentine brace failures and 15 other pectus carinatum patients who failed other forms of bracing, refused, or were unlikely to succeed with bracing were treated surgically. Thirteen underwent Abramson’s minimally invasive operation and 5 had an open repair, all with good initial correction. Open repairs were typically performed in the setting of significant chest wall stiffness in association with asymmetric, unilateral pectus carinatum and sternal torsion. For Abramson repairs, seven patients had bars removed with results graded as excellent (n = 4) or good (n = 2), with one failure (converted to open with excellent result). Wire breakage occurred in two of the first five patients, causing us to switch to flexible Mersilene tape to secure the bar to the ribs. Mersilene failed the six times it was used (including revising the two broken wires). We then switched to surgical cable, but this cut through the ribs in two patients (Fig. 4). In the last two patients we have used a washer-like rib protector (Fig. 5) and have had no further problems with attachment of the bar to the ribs. In three patients with stiff chests, we have resected costal cartilage thoracoscopically [12], measurably reducing the force needed to flatten the sternum.

3. Discussion

Haje’s 1979 report [6] brought brace treatment for carinatum to attention in modern times, but it may even have been used in the Middle Ages! An excavation below Ripon Cathedral in North Yorkshire, England, uncovered a 15th century young adult female. A photograph of her sternum very clearly shows pectus carinatum. There were flattening of the spinous processes of the third through the ninth thoracic vertebrae (T3–T9), and anterior flattening of the rib cage, raising the question of external compression [13].

The Argentine brace (Dynamic Compression System, or DCS) has given good results in our experience. Among its advantages over other orthotics is objective pressure measurement to guide treatment decisions. In addition, because the position of the compression plate is easily adjusted on its aluminum frame, the DCS facilitates the treatment of asymmetric deformities. We strongly endorse bracing as first-line treatment for patients with pectus carinatum. In our health care environment in the United States, surgical correction is considered only in those who fail bracing (11% of our group), refuse or show non-compliance with bracing, or who are unlikely to succeed with bracing due to chest wall stiffness or severe asymmetry.

Our data predict that flattening of the sternum can be expected in 6 (range 1–24) months with an additional 5 (range 3–19) months of retainer mode wear for a successful result. However, we note that several patients wore the brace for fewer hours than were prescribed during these months, e.g., wearing only during sleep, or only at school. Thus the question remains — how long does the adolescent need to wear the brace every day? It may be that the total duration of brace treatment can be shortened by increased daily wear. However, our data cannot resolve this question at this time.

These data suggest that there may be an initial pressure of correction that predicts brace failure. Trying to guide surgical decision-making, Martinez-Ferro et al. in review of their series, found that the brace failed in patients with an initial pressure of correction of more than 7.5 PSI [8]. We found a number of brace failures at lower correction pressures; the 13 failure patients had a median initial POC of only 5.5 (range 2.0–9.5) PSI. To further confuse matters, we have had a single patient (only one!) with a bracing success with an initial POC of over 9 PSI. Thus, our population can neither support nor refute the 7.5 PSI guideline as a threshold for predicting failure at this time, and we attempt bracing even with high pressure patients. It has been helpful, in this situation, to explain to the patient and family at the time of
initial consultation that total active treatment time would be expected to be longer and the chance of bracing failure higher with elevated POC. Even if a patient with a high initial pressure of correction fails bracing treatment, we have observed that the bracing attempt benefits the patient because it softens the chest, which helps surgical correction. Therefore, our clinical experience suggests that it is worthwhile to attempt bracing even with patients with high initial POC.

Because of the success of bracing treatment, the patients undergoing surgery in our case series represent a selected population, more difficult cases, with stiffer chest walls and more asymmetrical anatomy. The difficulties we have experienced in our Abramson cohort with breakage of wire and Mersilene tape may originate from the more inflexible nature of their chests as compared to those reported by Abramson [1,9]. But wire breakage might also result from the repeated bending which occurs with the 16,000+ respirations humans perform every day. Cable appears to provide the strength and flexibility needed for correcting these stiff chests. We posit that the constant friction of the cable on the posterior of the rib can lead to the cable cutting through the rib, akin to the action of a seton. The addition of a custom-designed rib protector, which acts like a washer to distribute force to the posterior rib appears to have eliminated this problem for our patients to date. In addition, thoracoscopic resection of the costal cartilages measurably decreases the pressure exerted on the bar by the chest and appears to aid in the success of the Abramson procedure. Intraoperative use of the DCS pressure device shows this phenomenon clearly.

Here, in our health care environment, we have pursued first-line treatment for pectus carinatum with the DCS brace with good results. Surgical treatment has been reserved for those who fail or are non-compliant with bracing therapy and those with chest wall stiffness or asymmetry that makes bracing unlikely to succeed. Even for patients who fail bracing, this treatment may assist in softening the chest wall in preparation for the Abramson procedure. For our Abramson cases, we secure the bar and stabilizer with cable and a rib protector washer to provide both strength and protection to the rib. Additionally, we have found that modifying the Abramson repair to include thoracoscopic resection of the costal cartilages can significantly increase compliance for patients with stiff chests or difficult chest wall anatomy.

**Acknowledgment**

We gratefully acknowledge the efforts of the following individuals: Marcelo Martinez-Ferro, M.D (guidance with brace treatment); John Duperock, MSPT, April Kirkner, DPT, Kimberly Kranz, PT DScPT SCS, and Sara Rau, DPT (dedicated physical therapy for our bracing patients); Craig Goodmurphy PhD (photographic assistance); and Trisha Arnel (secretarial assistance).

**References**