Nonoperative management of pectus carinatum


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Abstract

Background: Although surgery has been the mainstay of treatment of chondrogladiolar pectus carinatum (PC), several authors have advocated the benefits of nonoperative approaches to induce chest wall remodeling. Based on our initial success with compression bracing, we have integrated this modality into our treatment algorithm.

Method: We reviewed the charts of all patients treated for PC at our pediatric hospital between 1997 and 2004. Patients were managed with observation, operative repair, and orthotic bracing that provides continuous anteroposterior sternal compression. The brace was worn for 14 to 16 hours per day until linear growth was complete or for a minimum of 2 years.

Results: One hundred patients were diagnosed with PC. Fifty-seven patients had no treatment and were monitored. Twenty-nine patients were fitted with a brace. Of these 29 patients, 3 were noncompliant, resulting in a compliance rate of 90%. Of the remaining brace patients, all have had positive outcomes with no observed complications. Seventeen patients underwent surgical repair. Their outcomes were also positive with no major complications.

Conclusion: Our findings clearly demonstrate that compression bracing is a safe and effective treatment for children with chondrogladiolar PC. We currently offer this approach as a first-line treatment, reserving surgery for patients who are noncompliant and those who fail the nonoperative modality.

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1. Background

Pectus carinatum (PC) comprises a spectrum of anterior chest wall deformities characterized by convex protrusion of the sternum and adjacent costal cartilages. These deformities are classified as either chondrogladiolar or chondromanubrial, depending on the anatomical site of greatest prominence. Chondrogladiolar deformity is the symmetric or asymmetric protrusion of the gladiolus and inferior costal cartilages. In the asymmetric subtype, unilateral overgrowth of the costal cartilages results in a rotational deformity of the sternum and a keel-like appearance of that side of the chest. Chondromanubrial PC is the protrusion of the manubrium and superior costal cartilages; this subtype represents less than 1% of PC cases [1]. No definitive etiology of PC has been established; however, the increased incidence of positive family history (25%) of chest wall deformity suggests a genetic linkage [2]. Pectus carinatum also occurs in association with scoliosis (15%), congenital heart disease, Marfan syndrome, and other connective tissue disorders [2]. Pectus carinatum...
occurs more frequently in males than in females (4:1 ratio) [2]. It often progresses in severity and becomes quite prominent during the active linear growth spurt of puberty, causing significant cosmetic and psychosocial concerns. In addition, children frequently report symptoms such as chest pain and shortness of breath.

Surgery has been the mainstay of PC treatment for the past 50 years, and during this time, a number of technical and management modifications [3-6], including optimal timing of the procedure [5], have been advocated. Despite the success of operative approaches, the risks associated with any major surgical procedure remain. [7] Based on the fact that the anterior chest wall is still compliant during puberty, several authors have alternatively suggested a variety of nonoperative approaches to induce chest wall remodeling in patients with chondrogladiolar PC. Vidal et al [8] reported limited success with the use of a plaster cast followed by a plaster jacket. Mielke and Winter [9] reported a successful cosmetic outcome in a 14-year-old girl managed with an underarm body cast followed by bracing. Both Haje and Bowen [10] and Egan et al [11] have reported positive outcomes with a custom-fitted compressive orthotic. Encouraged by the senior author’s (RGA) initial experience with a similar nonoperative approach, we have integrated the use of an orthotic brace during puberty into our treatment algorithm. This report describes our 7-year experience.

2. Patients and methods

We reviewed the charts of all patients treated for PC at Cincinnati Children’s Hospital Medical Center between 1997 and 2004. Data pertaining to demographics, pertinent medical history, PC management, complications, and clinical outcome were obtained. This study was approved by our institutional review board.

2.1. Patient management

All patients initially underwent a thorough physical examination. This was followed by a variable period of observation to monitor PC progression and the consequent need for intervention. Decisions as to whether to initiate bracing as opposed to surgery were dependent upon differences in faculty practice and experience. As long as the chest wall of patients was assessed to be compliant by the treating surgeon (patients up to age 16), compression bracing was considered as a treatment option. When surgical repair was performed, a modified Ravitch procedure was used. Resolution or improvement of PC was determined by sequential physical examinations, pretreatment and post-treatment photographs, and patients’ own accounts of change in their chest wall contour.

2.2. Orthotic brace

Through collaboration with a certified orthotist, we designed a custom-fitted chest compression brace that is easily concealed under clothing. The brace consists of separate anterior and posterior compression plates that are anchored to aluminum struts. The struts of each compression plate are bound together by an adjustable leather strap on each side. The brace is constructed so to apply continuous anterior-posterior chest compression, pushing the sternum with the midback serving as a stable base (Fig. 1A and B). Patients are instructed to wear the brace over a T-shirt for 14 to 16 hours per day and to symmetrically tighten the leather straps as tolerated. Initially, the straps are tightened every 2 to 4 weeks. This is followed by adjustment tightening at less frequent intervals. After the initial brace fitting, patients are seen at 6 weeks to 3 months and then at 6- to 12-month intervals. Pressure on the anteroposterior projection of the chest is easily monitored; when pressure is sufficient, a red mark over the area of sternal protrusion persists for several hours after brace removal. Patients and families are informed that the brace is to be worn until linear growth is complete or for a minimum of 2 years.

3. Results

Between 1997 and 2004, a total of 100 patients (80 males, 20 females) seen at the Cincinnati Children’s Hospital were...
diagnosed with PC. The mean age of males was 13.1 ± 3.4 years (range, 1-18 years), whereas that of females was 11.7 ± 3.3 years (range, 6-17 years).

Of 100 patients, 57 had no specific treatment and were monitored over the duration of the study interval. During the course of the study, 29 of 100 patients were fitted with a brace as their treatment. Of these 29 patients, 3 were noncompliant. In 1 patient, noncompliance was because of behavioral problems. In another, it was because of developmental delay and the resultant lack of comprehension. In a third patient, noncompliance was caused by parental anxiety concerning the effect of bracing on breast development. There was thus a 90% brace compliance rate. The mean age of the remaining 26 patients was 12.4 ± 2.7 years (range, 6-16 years). These patients wore the bracing device for a mean period of 2.5 years (range, 1-3 years). Most patients removed the brace at nighttime to sleep. With the exception of 3 patients, all wore the brace to school and during athletic activities; however, these 3 readily adapted to wearing the brace after school and at night. Of the 26 brace wearers, 4 have not been followed long enough to determine outcome but are, however, showing promising initial results. All of the 22 patients who have been followed for more than 6 months have shown either significant improvement or complete resolution of their deformity (Fig. 2A, B and Fig. 3A, B). Some showed improvement as early as 6 weeks. In most patients, significant correction was observed within the first year and there appeared to be a plateau in improvement after 18 months. As expected, patients aged 12 to 14 years had a more rapid resolution of PC than those aged 14 to 16 years. One 8-year-old boy within this group developed PC after a Nuss procedure for pectus excavatum. One year after this procedure, the substernal bar was removed and he was then successfully treated with a brace.

Although persistent red marks occurred at primary brace pressure points during treatment, there was no evidence of skin breakdown. No complications were observed, and patients who had previously experienced chest pain or pressure or shortness of breath with exertion were asymptomatic after treatment. As evidenced by the 90% compliance rate as well as by verbal affirmation by patients and their families, overall satisfaction with bracing was high. Although compliant, 1 patient commented that it was difficult to wear the brace during summer in that it was uncomfortably hot.

Of 100 patients, 17 (14 males, 3 females) underwent surgical correction. Of these 17 patients, 3 had begun treatment with bracing but were unable to comply with this approach. The mean age at surgery was 15.0 ± 1.5 years (range, 11-18 years). Outcomes were successful, and no major complications occurred. Within this group, 1 patient also developed PC after a Nuss repair for pectus excavatum and had a modified Ravitch open repair to correct the PC. Bracing was not offered to this patient.

![Fig. 2](Image) A, Female patient with marked chondrogladiolar PC before bracing. B, A female patient after 1.5 years of bracing.

![Fig. 3](Image) A, Male patient with an asymmetric chondrogladiolar PC before bracing. B, The patient after 1 year of bracing.
4. Discussion

Our findings clearly demonstrate that our nonoperative approach to the treatment of chondrogladiolar PC in children is safe, effective, and without any apparent risk for adverse sequelae. Using an orthotic device to reposition the sternum and costal cartilages, we successfully managed 26 children with chondrogladiolar PC. Over the study interval, all achieved either significant improvement or complete resolution of their deformity by wearing an unobtrusive custom-fitted brace under their clothing. The brace was worn for 14 to 16 hours per day over an average period of 2.5 years. It is constructed so to apply continuous anteroposterior compression on the compliant chest wall. No complications were associated with bracing, and although one might expect difficulties with compliance in adolescent patients, our results showed a remarkably high (90%) compliance rate. Patients and their parents were especially pleased with a treatment option that averted major surgery and hospitalization. In addition, nonoperative management offers a significant cost benefit both to patients and third-party payers. The combined charges for hospitalization and professional fees for PC surgical repair in our institution exceeds $20,000, whereas the charges for the brace and outpatient visits are approximately $700. This is a particularly important advantage for bracing because, despite the associated physical symptoms of PC and the serious impact it often has on self-image, many health insurance companies deny coverage for corrective PC surgery, considering it to be primarily cosmetic.

The established record of corrective PC surgical procedures does not obviate the risks associated with any major surgical procedure. Although infrequent, complications such as pneumothorax, bleeding, hypertrophic scarring, wound infection, seromas, and pleural effusion still remain [7]. Also, females in early puberty have been reported to have mammary hypoplasia after PC repair [12]. In an effort to eliminate these risks, several authors have attempted less invasive, nonoperative approaches, using the known response of bone and cartilage to external physical force. Haje and Bowen [10] have advocated the use of a device called a dynamic chest compressor, which is constructed of wooden pads and U-shaped metal rods. Patients in this study were advised to wear the device 23 hours a day for 6 months for the first 6 months of treatment and instructed to perform deep breathing exercises for 20 minutes, 3 times daily. Egan et al [11] reported modest improvement in 5 teenage boys with asymmetric chondrogladiolar PC who wore a similar device. These authors used a radiographic marker to monitor initial sternal rotation and subsequent changes with growth or treatment. Although patients in our series wore their brace for a shorter period daily (14-16 hours), successful outcomes were achieved, with children at a stage of active vertical growth showing a quicker response to treatment than those who had completed vertical growth. Overall, our success attests to the importance of patient compliance and motivation, as well as supportive parents.

Unlike patients with chondrogladiolar PC, those with chondromanubrial PC are best treated by surgical intervention. In this PC subtype, there is an abnormal fusion of multiple rib cartilages at the manubri- gladiolar junction (third-fifth). This region is unlikely to respond to bracing because of these abnormal noncompliant chest wall structures. Because corrective surgical repair in all patients younger than 13 years with chondrogladiolar PC is associated with a 5% to 10% recurrence rate [7], in our clinical practice, we prefer to delay surgery until midadolescence.

Based on our positive results with the use of orthotic bracing, we are currently offering this modality as a first-line treatment for patients with chondrogladiolar PC and reserving corrective surgery for patients who are noncompliant or those who have failed with this nonoperative approach.

References

Discussion

Donald Nuss, MD (Norfolk, VA): I want to congratulate you on a very fine study and your outstanding results. There is no question that the chest is malleable. We children who suffer from neurologic impairment and cerebral palsy who start off life with a normal looking body and, at the end of a terrible time of cerebral palsy, end up with major chest deformity, so we know the chest can change because of various forces acting on it. It therefore makes sense that one should be able to correct some of these deformities, and you have shown an excellent result. We have also tried it except our brace is not as good as yours, and we have had some successes but not as high a percentage as you have. I have also spoken to Dr Martinez Faro(?) in Buenos Aires. They see a very large number of pectus carinatum. In fact, they see more carinatums than excavatums, and they have developed a brace very similar to yours, and they have been very happy with their results as well.

Ala Stanford Frey, MD (response): Thank you.

Marshall Schwartz, MD (Philadelphia, PA): I think this is a very interesting study. You stated that out of your 100 patients, 29 patients wore the brace. What about the other 71 patients? Did these patients or families decide not to wear the brace? Did you select the patients that wore the brace based on their likely commitment to continue to wear the brace? As you alluded to, teenagers are not particularly compliant. Wearing a chest brace for 14 to 16 hours a day for a prolonged period of time is a huge commitment. We have a hard enough time getting teenagers to take necessary medication, so how did you select and/or motivate these patients to stick to the commitment to wear the brace?

Ala Stanford Frey, MD (response): Thank you, Dr Schwartz. First, this was retrospective. Of the 100, 57 patients had no further intervention. Most of those defects were mild, and in the earlier patients, the only option we had for those patients was operative and most of the families and the children decided that they did not want to have an operative correction because their defect was not as prominent. Seventeen of the patients did undergo surgical correction. Three of our patients who were in the brace group went on to surgical correction because they were noncompliant. The one did not understand with his developmental delay. The second we could not stop him from smoking let alone wearing a brace, and so his parents opted that he should have a surgical correction. And the last, a mother was concerned about the breast development so still most of our patients with a mean age of 12 were compliant. We did not select the ones that we thought would be more compliant for the brace because this was retrospective and the fact that the varying staff, depending on the age of the patient, for example, if they were 16 or 17, their chest wall being less compliant, were less likely to benefit from brace compression. Therefore, they have a modified Ravitch repair.

Eric W. Fonkalsrud, MD (Santa Monica, CA): I would like to congratulate you and your colleagues from Cincinnati on this very innovative and very well presented group of patients. The question I have is that the average age of your patients at the time of application was about 12 years, I believe you said. The vast majority of patients with pectus carinatum just begin their protrusion during the early adolescent growth years and this progresses throughout the adolescent years up to about age 18 or 19. The question is, have you followed any of these patients during this adolescent growth period from say 13 to 16 when the patient is likely to grow 5 to 7 inches in height and the chest wall tends to become much more protuberant. Are there recurrences? Do you have to reapply the brace in any of those patients?

Ala Stanford Frey, MD (response): Thank you, Dr Fonkalsrud. The longest follow-up that we be actually a 7-year period, and those patients have not required an operation. Some may have required a reapplication of the brace but have not required intervention, ie, surgical intervention, to repair. The mean age was 12.4 with our standard deviation of ±3. The majority of the girls were 12 and the boys 13 through 16, as you mentioned, and, yes, they do with their linear growth have more protrusion; however, we fortunately have not seen recurrences with the brace in our population.

Jonathan Greenfeld, MD (Tucson, AZ): Two quick questions. First, did you grade the severity of the carinatum defects? The first picture you showed at the beginning of your presentation showed a very severe defect. Subsequent pictures, I think many of us would consider mild or moderate defects. Second, in follow-up to Dr Fonkalsrud, we have occasion to see older patients who do not have access to medical care earlier in their years who present in the late adolescent period. Dr Nuss has noted that even these patients will have malleable chest wall. Have you tried the brace in those patients?

Thank you.
Ala Stanford Frey, MD (response): In answer to your second question first, in the patients that are 16, 17 years of age, their chest wall, yes, is less compliant. I do not think that you lose anything by giving them a trial with the brace first, knowing that their success with the brace is probably going to be less than the younger age patients that have not completed their growth.

Your other question about access to health care and so forth, that those patients present later, we have actually found that, in using the brace, the cost of the brace, the follow-up visits, if this were offered, is $700 as opposed to over $20 000 for an open repair in patients, so if they were noticed by a pediatrician, for example, and they knew at your institution that this was available, this would be something that could be offered.

Your first question last about the severity index, we contemplated measuring the transverse by anterior posterior diameter. However, in our patients, since the majority of them were asymptomatic save for some having chest pain for the most part that was relieved by NSAIDs and ice packs, we did not feel that it added anything objectively to our study because the patients could tell us by looking in the mirror that they felt better, and that was the most accurate measurement of improvement and success with the brace.