Improvement of cardiopulmonary function after minimally invasive surgical repair of pectus excavatum (Nuss procedure) in children

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ABSTRACT

Background : **Severe pectus excavatum in children may result in cardiorespiratory functional impairment; therefore, we evaluated cardiopulmonary response to exercise before and after the Nuss procedure.**

Methods : **Twenty-four physically active pediatric patients aged 9–18 years with severe pectus excavatum (Haller index >3.25) were included in the study. Cardiopulmonary exercise testing using treadmill and modified Bruce protocol was performed before and after the Nuss procedure.**

Results : **Maximal oxygen uptake and oxygen pulse improved by 40.6% (32 ± 13–45 ± 10 ml/kg/min; $P = 0.0001$) and 44.4% (9 ± 4–13 ± 5 ml/beat; $P = 0.03$), respectively, after surgical correction of pectus excavatum by Nuss procedure. Significant improvement in maximum voluntary ventilation and minute ventilation after Nuss procedure was also noted.**

Conclusions : **We found that, after repair of pectus excavatum by Nuss procedure, the exercise capacity as measured by maximal oxygen consumption improved significantly primarily due to increase in oxygen pulse, an indirect measurement of stroke volume.**

Keywords : Cardiopulmonary exercise stress test, Haller index, Nuss procedure, pectus excavatum

INTRODUCTION

Pectus excavatum, also known as funnel chest or sunken chest, is by far the most common congenital chest wall deformity, occurring in approximately one in 400 births and in males more frequently than in females.[1] It is more common in Caucasians. Although the etiology is not fully understood, it is hypothesized that pectus excavatum occurs due to structural abnormality of mesenchyme and results in unbalanced overgrowth in the costochondral regions that leads to a concave appearance of the anterior chest wall. One study showed that asymmetric pectus excavatum patients had shorter ribs on the more severely depressed side of the deformity.[2] It is hereditary, but neither the exact mode of transmission nor the exact gene is known; up to 40% of pectus excavatum patients have a family history of the disease.[3] Pectus excavatum can be associated with scoliosis and connective tissue disorders such as Marfan syndrome, Ehlers–Danlos syndrome, and Noonan's syndrome. Although pectus excavatum is usually present since birth, most notice development and significant progression of the deformity during puberty. This may be accompanied by onset or worsening of symptoms.

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Symptoms may include disproportional dyspnea on exertion, decreased exercise endurance, exertional and nonexertional chest pain, back pain, progressive fatigue, palpitations, exercise-induced wheezing, frequent respiratory tract infections, and syncope/presyncope. Chest wall restriction and decreased thoracic volume are believed to play a role in decreased filling of both ventricles. Cardiac compression, especially the right ventricle, has been shown to reduce left ventricular (LV) ejection fraction, with improvement after pectus excavatum repair.[4] The psychosocial issues associated with altered body image and self-esteem are often significant, especially in adolescents and young adults. The importance of "cosmetic concerns" to affected individuals and their families should not be underestimated by pediatricians. In summary, pectus excavatum is not only a cosmetic problem but also a functional issue that can alter cardiopulmonary physiology.

We studied the effect of pectus excavatum on cardiopulmonary response to exercise before and after repair, taking into consideration the severity of pectus excavatum (Haller index by computed tomography [CT] scan) to determine if there was any improvement in cardiopulmonary function.

METHODS

Patients

Patients with pectus excavatum, who were referred for surgical correction, underwent a complete evaluation to rule out other chest wall deformities such as pectus carinatum, scoliosis, and Marfan syndrome. Habitual exercise activity was assessed for each patient in terms of frequency and duration of exercise participation. All patients had an echocardiogram, electrocardiogram, chest roentgenogram, and CT scan of chest. CT scans were used to determine the severity of the pectus deformity (Haller index).[5] The Nuss procedure (minimally invasive repair of pectus excavatum)[6] was used to repair the pectus excavatum if patients demonstrated any two or more out of the four criteria listed in Table 1. Video-assisted thoracoscopy was used to facilitate insertion of a custom-bent curved metal bar underneath the sternum through a lateral chest wall incision. The bar was then turned, placing the convexity of the bar upward correcting pectus excavatum. No resection of cartilage was performed. The bar was then secured to the lateral aspect of the chest wall, ribs, or both and left in place for a minimum of 2 years. Repeat cardiopulmonary exercise and pulmonary function tests were performed 1–2 years after bar insertion, with the bar in place, after patients were fully recovered from the surgical procedure. Institutional review board approval was obtained for retrospective analysis of medical record of all patients for this study.

Table 1: Criteria for surgical repair of pectus excavatum

Chest wall deformity as noted by Haller index *>3.25
Evidence of cardiopulmonary limitations (any one or more of the following)
Decreased maximum oxygen utilization
Restrictive pulmonary disease
Right-sided cardiac compression seen by echocardiogram or chest CT
Shift of heart into left thorax
Any one or more of the symptoms: Disproportional dyspnea on exertion, decreased exercise endurance, exertional and nonexertional chest pain, back pain, progressive fatigue, palpitations, exercise-induced wheezing, frequent respiratory tract infections, and syncope/presyncope
Severe body image disturbances

*Haller index of severity was calculated by measuring the inner width of the chest at the lowest level of pectus excavatum defect and dividing it by the anteroposterior distance (between the posterior surface of the sternum and the anterior surface of the spine). CT: Computed tomography

Cardiopulmonary exercise testing

All patients before surgery underwent cardiopulmonary exercise tests utilizing treadmill per Bruce protocol, and respiratory gases were obtained from breath-by-breath analysis using a metabolic cart (V_{max} Encore, Viasys Healthcare, Yorba Linda, CA, USA) as recommended in the guidelines of clinical stress testing in the pediatric age group published by the American Heart Association.[7] Each patient underwent baseline spirometry followed by a progressive cardiopulmonary exercise with continuous monitoring of electrocardiogram, ventilation, oxygen saturation, gas exchange, and blood pressure. The patients were strongly encouraged to exercise to the point of exhaustion. Tests suitable for analyses reflected maximal effort based on an achieved respiratory quotient of >1.0.[7]

The parameters of exercise performance such as maximum oxygen consumption (ml/kg/min); oxygen consumption at ventilatory threshold (ml/kg/min); ventilatory efficiency as measured by the rate of rise in ventilation per amount of expired carbon dioxide below the respiratory compensation point; oxygen pulse (ml/beat), defined as oxygen consumption divided by heart rate; resting and peak heart rate (beats/min);, heart rate reserve (peak-predicted heart rate of 210 – age – peak heart rate attained); resting and peak systolic and diastolic blood pressures; and oxygen saturation at rest and at peak exercise were measured.[7] Peak oxygen consumption was taken as the maximum oxygen consumption value obtained. The percentage of predicted peak oxygen consumption is calculated using previously established normal values.[8] Anaerobic threshold was determined using the "V-slope" method.[9] The minute ventilation/pulmonary carbon dioxide output slope was determined and used as a measure of ventilatory efficiency.[8] Age and gender normative data were used for determining percentage-predicted values.[8]

Pulmonary function testing

Spirometry was performed in the seated position with a nose clip that was applied after the patient rested for at least 10 min. Testing was performed by a respiratory therapist using equipment and procedure that meet the American Thoracic Society criteria for standardization of spirometry.[10,11] Because pulmonary function tests are effort dependent, patient performed several trials with strong verbal encouragement from the respiratory therapist. Parameters for pulmonary functions such as forced expiratory volume in 1 s, forced vital capacity, Tiffeneau–Pinelli index (forced expiratory volume in 1 s/forced vital capacity), and maximum voluntary ventilation (MVV) measurements were obtained at rest before cardiopulmonary exercise test. Maximum minute ventilation and ventilatory reserve, i.e., percentage, were calculated as previously described.[8] The pulmonary function test results were expressed as measured values and as percentages of gender- and age-specific reference values.[12]

Statistical analysis

Cardiovascular, ventilatory, and gas exchange data were presented as mean and standard deviation. Demographic and cardiopulmonary variable differences before and after Nuss procedure cohorts were assessed using unpaired *t*-test. Data were also presented as percentage of reference value as appropriate. The means and quartiles for maximum oxygen consumption and oxygen pulse were presented as boxplot with maximum and minimum values. $P < 0.05$ was considered statistically significant.

RESULTS

Our sample consisted of 24 physically active pectus excavatum patients, aged 9–18 years, who underwent cardiopulmonary exercise tests and spirometry before and after Nuss procedure. The echocardiogram did not show any specific anatomical or functional abnormality, and none of the patients have direct or indirect evidence of pulmonary hypertension. All patients included in the study were physically active, as defined by moderate regular aerobic activity for a duration ranging from 30 min to 2 h/day for ≥3 days per week.[13] As shown in Table 2, there was no statistically significant difference in patients' age, height, and weight at the time of pre- and post-Nuss cardiopulmonary exercise tests. Pectus severity (Haller index) as estimated by CT scan was 4.3 ± 0.9 (range: 3.05–6.64). There was no difference in Haller index calculated by CT scan and chest roentgenogram ($P = 0.8$).

Cardiovascular response to exercise before and after Nuss procedure

Figure 1 demonstrates that, before surgical repair, pectus excavatum patients have decreased maximum

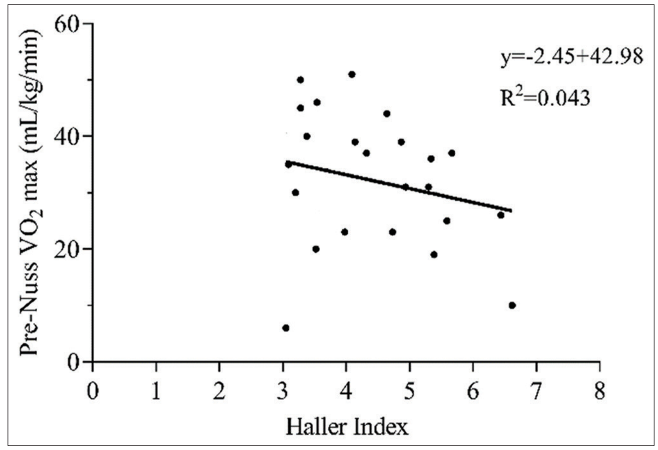

Figure 1: A negative linear relationship between maximum oxygen utilization and severity of pectus excavatum (Haller index)

oxygen consumption which correlated inversely to pectus severity. The maximum heart rate (increased by 12.4%), percentage-predicted maximum oxygen consumption (increased by 41.9%), and percentage-predicted oxygen pulse (increased by 26.3%) after Nuss procedure were statistically significantly higher than before (Nuss) repair as shown in Table 3. Maximal oxygen uptake and oxygen pulse were improved by 40.6% (32 ± 13–45 ± 10 ml/kg/min; $P = 0.0001$) and 44.4% (9 ± 4–13 ± 5 ml/beat; $P = 0.03$), respectively, after surgical correction of pectus excavatum by Nuss procedure [Figure 2]. The box-and-whisker plots in Figure 2a and b described the mean, the maximum and minimum values, and quartiles of the changes in maximum oxygen consumption and oxygen pulse before and after Nuss procedure.

Ventilatory response before and after Nuss procedure

Static pulmonary function indices such as forced expiratory volume in 1 s, forced vital capacity, and Tiffeneau–Pinelli index at rest before Nuss procedure were in the normal range compared to the value predicted by the patient's age, height, weight, and gender as shown in Table 3. Therefore, as expected, there was no significant change in these parameters after Nuss procedure in resting condition except percentage-predicted forced expiratory volume in 1 s, which increased by 5.8%, $P < 0.0003$ [Table 3]. To evaluate whether the patient exhibited ventilatory limitation to exercise, we compared MVV and ventilatory reserve, i.e., percentage-predicted MVV at maximum exercise before and after Nuss procedure. As shown in Table 3, there was a significant increase in MVV (32.9%, $P = 0.01$) and maximum minute ventilation (40.6%, $P = 0.01$) after Nuss procedure. However, there was no significant change in ventilatory reserve after Nuss procedure compared to that before the repair of pectus excavatum.

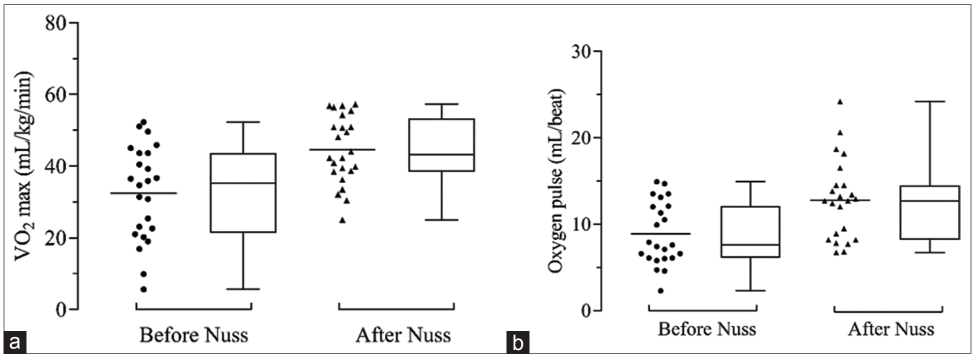

Figure 2: The mean change in maximum oxygen utilization (ml/kg/min) (a) and oxygen pulse (ml/beat) (b) at exercise before and after Nuss procedure with means on the left and quartiles with maximum and minimum of all values on the right

Table 2: Demographic data of patients with pectus excavatum

	Before Nuss procedure	After Nuss procedure	*P*
Age (years)	12.9±3.6	14.6±3.7	0.1
Weight (kg)	45.5±16.8	54.4±17.9	0.1
Height (cm)	157±22.2	165±19.3	0.2
BSA (m^2)	1.4±0.4	1.6±0.4	0.1
Haller index by CXR	4.4±0.9*	2.5±0.25	<0.001
Haller index by CT	4.3±0.9*		

*There is no difference between Haller index estimated from chest roentgenogram and CT scan (*P*=0.8). CT: Computed tomography, BSA: Body surface area, CXR: Chest radiography

Table 3: Cardiovascular, ventilatory, and gas exchange responses to exercise before and after Nuss procedure

Parameters	Before Nuss procedure	After Nuss procedure	*P*
Cardiovascular, ventilatory, and gas exchange responses to exercise			
Resting heart rate	80±14	74±11	0.09
Maximum heart rate bpm	169±29	190±15	0.002
VO_{2max} (ml/kg/min)	32±13	45±10	0.0001
Percentage-predicted VO_2	62±25	88±16	0.0005
Oxygen pulse (ml/beat)	9±4	13±5	0.03
Percentage-predicted oxygen pulse	76±24	96±16	0.001
VE/VCO_2	30±5	30±4	0.5
MVV	91±32	121±41	0.01
VEmax	64±25	90±41	0.01
Percentage MVV	69±21	71±16	0.7
Resting PFT			
FEV_1	2.65±0.9	3.1±1.04	0.12
Percentage-predicted FEV_1	85±11	90±15	<0.0003
FVC	3.2±1.1	3.7±1.2	0.14
Percentage-predicted FVC	85±13	92±13	0.13
FEV_1/FVC	80±6	80±11	0.8

FEV_1: Forced expiratory volume in 1 s, FVC: Forced vital capacity, PFT: Pulmonary function test, MVV: Maximum voluntary ventilation, VE: Minute ventilation, VCO2: Pulmonary carbon dioxide output, VE_{max}: Maximal expired minute ventilation, VO_2: Oxygen uptake, VO_{2max}: Maximal oxygen uptake

Normalization of the chest wall deformity

Chest roentgenograms showed substantial normalization of the chest wall deformity with decrease in Haller index from 4.4 ± 0.9 to 2.5 ± 0.25 ($P < 0.001$) after the Nuss procedure.

DISCUSSION

This study investigated cardiopulmonary exercise test and pulmonary function parameters to assess the effect of pectus excavatum deformity on patients' cardiopulmonary function during exercise in a systematic manner. There is controversy about true physiologic alteration in pectus excavatum patients or mere reflection of deconditioning due to lack of physical activity or psychological discouragement from exercise participation. To address these issues, the current study examined the effect of pectus excavatum on ventilatory and cardiovascular responses to exercise in patients who engaged in moderate aerobic activity before the Nuss procedure, on an average 1 h/day, at least 3 days per week. There is 44.4% improvement in stroke volume (the surrogate marker is oxygen pulse) and 40.6% improvement in cardiac output (the surrogate marker is maximum oxygen utilization) after Nuss procedure.

In this study, we have shown that patients with severe pectus excavatum have lower maximum oxygen consumption before surgery. There is a correlation between maximum oxygen consumption and severity of pectus deformity as estimated by Haller index. A recent study has shown similar correlation between Haller index and decreased cardiac output.[14]

In our study, pectus excavatum patients have decreased oxygen pulse (surrogate marker for stroke volume) during exercise. Because of this, there occurs increase in heart rate, followed by a decrease in cardiac output, which leads to a limitation in peak exercise capacity as measured by maximum oxygen consumption. Increased severity of the pectus deformity then leads to a proportionately decrease in cardiac output.[14] Multiple studies have demonstrated that patients with pectus excavatum have reduced diastolic filling of their heart and low stroke volume.[15-19] A 19% increase in oxygen pulse after pectus repair was previously reported by Kelly *et al*.[20] Another study by Sigalet *et al*.[21] has shown that pectus excavatum patients have achieved higher oxygen pulse after Nuss surgery than before surgery although not quite as large as our current study. The improvement of maximum oxygen consumption in our study after surgical repair appears to be due to increase in stroke volume during exercise, and our results are similar to other published studies.[22]

The chest wall deformity due to pectus excavatum places the inspiratory and expiratory muscles as well as the rib cage in a mechanical disadvantage and pulmonary functional indices including forced expiratory volume in 1 s, forced vital capacity, and total lung capacity is reduced.[23] Malek *et al*. have reported their results of meta-analysis that cardiovascular function improves significantly, whereas pulmonary function does not significantly improve after surgical repair in pectus excavatum patients.[24]

In our study, there is little overall change in static pulmonary function indices, such as forced expiratory volume in 1 s, forced vital capacity, and Tiffeneau–Pinelli index before and after surgical repair contrary to findings of Kelly *et al*.,[20] who have shown that there is a significant improvement in lung function at rest after surgical correction of pectus excavatum. However, we have found higher percentage-predicted forced expiratory volume in 1 s after surgical repair in the present study. The improvement in lung function parameters is seen to some extent in those with the most severe degrees of pectus excavatum before surgery, but not statistically significant. Static pulmonary function test parameters are less sensitive in demonstrating compromised pulmonary function than dynamic pulmonary function testing parameters.

Among the dynamic lung function tests, MVV, percentage-predicted MVV, and maximum minute ventilation during exercise reflect a combination of chest wall compliance, muscular ability, and patient effort and are better parameters to evaluate the ventilatory limitation to exercise in pectus excavatum patients.[25] In our study, MVV and maximum minute ventilation are significantly improved after surgical repair; however, there is no statistically significant change in ventilatory reserve. This finding is not surprising as most of our patients had no significant limitation on their static pulmonary function test parameters at rest before the Nuss procedure.

The study limitations include (a) no echocardiographic study to assess right ventricular or LV function during surgical repair was available to confirm the change in stroke volume before and after Nuss procedure, and (b) because this is a retrospective study, magnetic resonance imaging was not a part of the study to routinely evaluate the impact of pectus excavatum on the cardiac function.

CONCLUSIONS

We conclude that, in physically active children with pectus excavatum, there is an inverse linear correlation between the exercise capacity as measured by maximum oxygen utilization and the severity of pectus deformity as determined by Haller index. In this study, cardiopulmonary exercise and dynamic pulmonary function tests parameters show a significant improvement in exercise capacity and corroborate previous studies that emphasize an increase in stroke volume during exercise after surgical repair of pectus excavatum using the Nuss procedure.

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Conflicts of interest

There are no conflicts of interest.

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