

Case report

EVALUATION OF UNEXPLAINED DYSPNEA IN A YOUNG ATHLETIC MALE WITH PECTUS EXCAVATUM

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ABSTRACT

Pectus excavatum (PE) is a relatively common congenital deformity of the anterior chest wall associated with reduced exercise capacity. Uncertainty exists over the nature of physiologic impairment in PE. Evidence suggests that myocardial compression exerted by the displaced sternum on the right heart chambers, disables the ability of the heart to augment stroke volume during exercise. This case study describes the evaluation of an athletic 20 year old Caucasian male, lifelong non-smoker, with severe pectus deformity and previous fixation procedure to repair a sternal fracture. The patient performed an incremental cycle ergometer exercise test to determine the etiology of his dyspnea with exertion. The patient demonstrated normal work output and normal aerobic capacity but displayed dynamic hyperinflation. Mechanical restriction of tidal volume expansion appeared to be the major contributors to exercise limitation. These results are compared and contrasted with similar cases reported in the literature.

KEY WORDS: Funnel chest, exercise test, ventilatory limitation.

INTRODUCTION

Pectus excavatum (PE) is a common congenital deformity of the anterior chest wall, which occurs in approximately 1 in 300 births, more frequently in male children by a 9:1 margin. The pectus deformity is characterized by an inward depression of the sternum that may be symmetrical, or asymmetrical, and may present with varied degrees of torsion of the sternum (Williams and Crabbe, 2003). It often worsens in late adolescence and early adulthood (Malek and Fonkalsrud, 2004) and there are reports of reduced exercise capacity (Shamberger, 2000).

This case study includes a description of the potential sources of exercise limitation in PE and provides an example of the clinical evaluation of unexplained dyspnea in a patient with pectus

excavatum. It discusses the similarities and differences between the current study and previous studies of patients with PE that observed cardiovascular and ventilatory responses at rest and during incremental cycle ergometry.

What is the source of exercise limitation in Pectus Excavatum?

There is wide debate whether PE causes limitation to exercise. Some authors contend that exercise limitations related to PE are medical myth. Other authors report non-significant differences for maximal workload, oxygen consumption, cardiac output, or stroke volume when patients with PE are compared to normal controls (Ghory et al., 1989; Haller et al. 1970). Other investigators report data to suggest that PE can unfavorably affect

cardiorespiratory function and reduce exercise capacity (Beiser et al., 1972; Cahill et al., 1984; Malek and Fonkalsrud, 2004; Peterson et al., 1985).

The literature offers one explanation that suggests posterior displacement of the sternum in PE can produce deformity of the myocardium with anterior indentation of the right ventricle (Garusi and D'Ettoire, 1964; Shamberger, 2000). The resulting compression of the right heart limits stroke volume augmentation during exercise (Haller et al., 1970). Several studies dating as early as 1960, have examined the impact of exercise performed in the supine and the seated position to confirm limitation in stroke volume in individuals with PE (Bevegård et al., 1960; Bevegård, 1962; Beiser et al., 1972; Gattiker et al., 1966; Zhao et al., 2000).

Another potential consequence of sternal displacement is rotation and translocation of the heart into the left thorax and is reportedly common in individuals with severe pectus deformity (Haller and Loughlin, 2000; Malek and Fonkalsrud, 2004; Williams and Crabbe, 2003). Malek and Fonkalsrud (2004) describe the leftward displacement of the heart in patients with PE as a palpable translocation of the myocardium to the left mid-axillary line slightly below the armpit. An illustration of this translocation is illustrated in Figure 1. The rotation and translocation of the heart could conceivably cause functional restrictive cardiomyopathy accompanied by torquing of the great vessels, which would also limit stroke volume augmentation. In such a scenario, increases in cardiac output would be constrained solely to increases of heart rate. Thus, one could expect a person with severe PE to maintain little heart rate reserve during vigorous physical exertion, which is consistent with "cardiovascular limitation" to exercise. Torquing of the displaced myocardium is evident in axis deviations observable on 12 lead ECG. However, this has not been studied to date. One additional factor with hemodynamic implications for the patient with PE is that co-existent mitral valve prolapse and PE is documented. This phenomenon is presumably due to deformation of the mitral annulus, a consequence of the anterior compression of the myocardium (Williams and Crabbe, 2003).

Other researchers have sought to determine if exercise limitation might be ventilatory in nature. PE is associated with restriction of lung volume, attributed by some authors to limitation of rib cage mobility. However, reductions in lung volume and rib cage mobility occur to a degree that should not adversely influence exercise tolerance (Gattiker and Buhlmann, 1966; Mead et al., 1985). Nevertheless, Morshuis et al. (1994) found ventilatory limitation occurred during exercise in 43% of their 35 participants, and accounts of patients with PE

reporting exercise limitation are common (Shamberger, 2000).



Figure 1. Illustration of pectus deformity and myocardial translocation. Pectus deformity with obvious leftward translocation of the myocardium which concurs with the description by Malek and Fonkalsrud (2004), and Garusi and D'Ettoire (1964) of translocation of the myocardium to the mid-axillary line slightly below the armpit. Scar at midline is due to repair of the sternal fracture, not pectus repair.

Some authors suggest the symptomatic impairment in PE is attributable to a decrease in intra-thoracic volume. However even healthy individuals demonstrate wide variability in pulmonary function which can be also dependent on physical conditioning. It can also in part be attributed to the tendency for patients with PE to slouch, thereby adversely influencing pulmonary function (Shamberger, 2000).

Orzalesi and Cook (1965) report the observation that in a cohort of 12 children with severe pectus that the group had significantly smaller vital capacity (VC), total lung capacity (TLC), and maximal breathing capacity compared with height matched normals. Cahill et al., (1984) also reported smaller vital capacities in their sample of 14 patients. Likewise, Weg et al. (1967) found in a group of 25 Air Force recruits who were tested based on respiratory symptoms and PE, that although there were no significant differences in mean vital capacities, maximal breathing capacity differed significantly from predicted normal values. Gattiker and Buhlmann (1966) found normal lung volumes as well, and only minimally reduced breathing capacity when their cohort of patients was compared to normals. Castile et al. (1982) reported mean total lung capacity was 79% predicted in a cohort of patients with PE. However, their seven patients did not exhibit flow volume characteristics that were suggestive of airway obstruction. Exercise testing revealed normal dead space/tidal volume

relationship (V_D/V_T) and did not reveal alveolar-arterial oxygen difference abnormalities which argues against significant ventilation-perfusion (V/Q) mismatching. The authors found however, that as the workload approached maximal, that the symptomatic patients exhibited “measured oxygen uptake that increasingly exceeded predicted values”, and at peak exercise, that VO_2 exceeded the predicted values by 25.4%, with vital capacity normal or only slightly reduced. The authors suggested that increased work of breathing might have been responsible for the increase in oxygen uptake.

However, a supposition that exercise limitation is ventilatory in nature is refuted by multiple reports that show normal ventilatory reserve (V_E/MVV of less than 0.70) in patients who had not undergone pectus repair (Wynn et al., 1990), and that physical improvement after pectus repair had not been explained by changes in cardiorespiratory function. Finally, other authors have stated that fatigue and reduced exercise tolerance in adolescent patients with PE have most likely been due to habitual inactivity (Williams and Crabbe, 2003).

Thus, there remains no consensus as to what degree, or the source of physiologic impairment that exists because of this chest deformity. Though the literature supports the source of exercise limitation as cardiovascular in nature, secondary to impairment of normal inotropic (Frank Starling effect) stroke volume augmentation.

Pectus Severity Index (PSI) as a clinical benchmark

It has been suggested that severity of pectus deformity is related to the exercise limitation. Therefore, it was thought that the use of computed tomography (CT) scans would be a useful tool to determine the severity of the pectus deformity. This led to the development of a Pectus Severity Index (PSI) (Haller et al., 1987), derived by dividing the internal width of the chest at the widest point, by the distance between the posterior surface of the sternum and the anterior surface of the spine. Whereas a mean index of 2.5 is considered normal, Williams and Crabbe (2003) reported a ratio of greater than 3.2 to be a benchmark for severe pectus deformity. Fonkalsrud et al. (2000) have observed symptomatic PE in patients with a severity index ranging from 3.2 to 12.78. The severity index has been reported to correlate to the predicted values of TLC and VC (Beiser et al., 1972). Malek et al. (2003) have stated that patients with a PSI of greater than 4.0 were also eight times more likely to demonstrate a reduction in aerobic capacity compared to patients with a lower PSI, despite their

level of exercise participation. The CT scan with thoracic dimension is displayed in Figure 2.

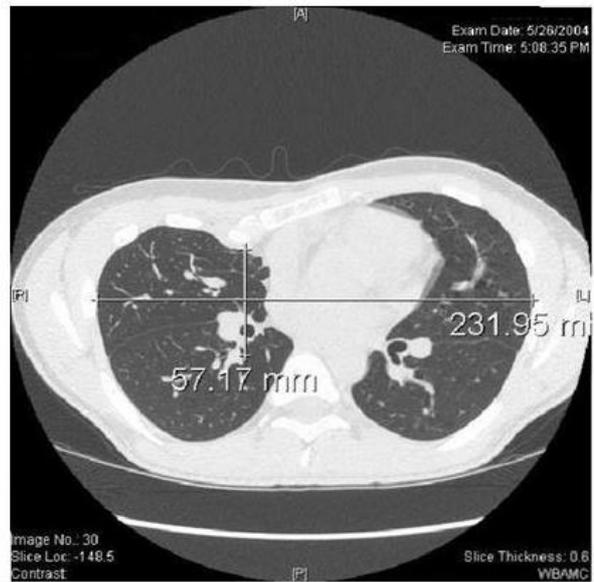


Figure 2. Computed Tomography Scan (CT) with thoracic dimensions. Digitally de-identified CT scan reveals pectus deformity. Pectus Severity Index (PSI) reveals an index of 4.06. A PSI of greater than 3.2 is considered to be a severe pectus deformity.

What role does cardiopulmonary exercise testing (CPET) play in this type of evaluation?

Cardiopulmonary exercise testing can be useful in a wide spectrum of clinical needs. In practice, it is useful in the clinical decision-making process including diagnosis, assessment of severity, monitor disease progression, prognosis, and response to treatment (American Thoracic Society / American College of Chest Physicians, 2003). The advent of widespread availability of computerized metabolic systems has permitted use of exercise testing in situations requiring differential diagnosis of exertional limitation due to not only cardiac factors, but related to ventilatory, gas exchange, musculoskeletal, or psychogenic factors as well. Since resting physiologic measures lack the reliability to predict exercise performance and functional capacity, there is a poor correlation between resting physiologic measurements and exertional symptoms. As such, the literature suggests that cardiopulmonary offers valuable insight regarding impairment of functional capacity, quantification of the factors limiting exercise, and the definition of the underlying etiology of exercise limitation such as the contributions of cardiac versus ventilatory factors. Frequently, the mechanism of exercise limitation is ventilatory in nature. As a result, techniques permitting the detection and grading of ventilatory limitation have become a practical tool in defining the source of unexplained exercise intolerance.

Historically, evaluation of the level of ventilatory limitation has been based on ventilatory reserve, or the degree to which peak minute ventilation (V_E) approaches measured maximal voluntary ventilation (MVV), or based on predictors of MVV such as FEV_1 multiplied by 35 or 40 (Beck, 1997). However, because the MVV test is characterized by short, high intensity effort performed in a breathing pattern that varies greatly from ventilation observed during exercise the test tends to overestimate ventilatory capacity.

The emerging clinical tool that provides unique clinical insight over these traditional measures of ventilatory limitation is the exercise tidal flow volume loop (extFVL). This technique provides a visual representation of the breathing pattern that allows the clinician to establish the degree of ventilatory limitation, and allows a more detailed approach to defining ventilatory limitation relative to the V_E /MVV relationship. In this regard, exercise flow-volume loops provide a non-invasive assessment of ventilatory mechanics, and permit a differential diagnosis not provided with traditional exercise testing. The extFVL also provides a determination of exercise inspiratory capacity (IC) that provides important clinical information regarding gas trapping.

What are the important Clinical Questions related to this case?

1. What was the source of this patient's unexplained dyspnea during exercise?
2. Did this patient exhibit a typical profile of exercise limitation for pectus excavatum?
3. Were there additional clinical considerations that may have affected the patient's exercise tolerance?

CASE REPORT

History

Our Human Performance Laboratory accepted a referral for a 20-year-old Caucasian male lifelong non-smoker with a congenital pectus excavatum deformity in order to determine the underlying mechanism for chronic dyspnea on moderate exertion. His occupation as a military policeman required him to wear body armor that reportedly exacerbated existing dyspnea. The patient, reportedly a former NCAA Division I basketball player had sustained a sternal fracture and the fracture of two ribs during a basketball game four years prior. He underwent a sternal fixation procedure to repair surgically the sternum at that time and since has maintained an active lifestyle. However, he has reported worsened dyspnea on exertion since arrival in El Paso, TX. There were no

reports of shortness of breath at rest, but the patient related that the dyspnea has always manifested as sharp left sided sub-costal pain at the mid-clavicular line with widespread radiation he described as "dullness". He reported occasional hemoptysis with extreme exertion. There were no reports of palpitations, syncope, nausea, vomiting, or diaphoresis. He stated that the pain has resolved between 30 to 240 minutes after termination of exertion. He reported no wheezing, but acknowledged a rare cough and chronic nasal congestion with postnasal drip.

Past medical history was significant for chronic bronchitis and gastroenteritis, mixed obstructive-restrictive pattern spirometry for which he has been treated, and a borderline positive methacholine challenge test ($PC_{20} = 6.7\text{mg}\cdot\text{ml}^{-1}$). Current medications included Advair 500/50, Montelukast (Singulair) 10mg, Fluticasone (Flonase) 0.05%, and Albuterol (Ventolin) 90mcg as needed.

Physical examination

Height 1.75 m and body weight 61.4 kg. Vital signs were normal. The patient was a thin, underweight male with obvious pectus deformity as illustrated in Figure 1. Vital signs; ears nose and throat; neck; and lymph nodes were all normal. Physical examination was significant for abnormal point of maximal impulse (PMI) clearly visible with displacement to the left, and pectus deformation of the sternum with a well-healed incision at the site of the sternal fixation procedure.

Laboratory findings

Hemoglobin (Hb) and hematocrit (HCT) were normal, as were creatine kinase (CK), creatine kinase-myoglobin (CK-Mb), and Troponin I.

Chest X-ray revealed hyper-expanded lungs that was corroborated by subsequent pulmonary function testing; residual volume (RV), 1.75L (123% predicted). Graded exercise test was normal with chest pain (5/10) 10 minutes into exercise, but no electrocardiographic changes were observed to support ischemia. Cardiac stress echocardiogram was normal with normal left ventricular ejection fraction (65%) and normal wall motion. The patient achieved 15 METS and a peak heart rate of 184 (92%), though the patient did report left sided pleuritic chest pain (5/10). Echocardiography with bubble study was performed with two injections of 10cc each of 0.9% sodium chloride to rule out a patent foramen ovale or other atrial/septal defect. Atria and left ventricle were normal size with normal wall motion. Left ventricular ejection fraction was low normal (55-60%) and aortic, mitral, pulmonic, and tricuspid valves were all normal. A helical computed tomography (CT) scan of the chest

showed normal lung parenchyma and a PSI of 4.06. A nuclear medicine lung perfusion scan was negative for pulmonary embolism. Spirometry and plethysmography were suggestive of obstructive-restrictive ventilatory impairment: forced vital capacity (FVC), 2.89L (52% predicted); forced expiratory volume in one second (FEV₁), 2.48L (53% predicted); FEV₁/FVC, 86%; total lung capacity (TLC), 4.65 (68% predicted); residual volume (RV), 1.75L (123% predicted). There were no significant changes post-bronchodilator. There was a moderate reduction in lung diffusing capacity (D_LCO adjusted), 5.4 mL·mHg⁻¹·min⁻¹ (68% predicted).

Pre-exercise spirometry and maximal breathing capacity

Prior to exercise, the patient performed forced spirometry and MVV according to the guidelines of the American Thoracic Society (1995). Spirometry and an estimate of maximal breathing capacity was obtained on a VMax Spectra mass flow sensor with a Free Flow mouthpiece and Micro Guard microbial filter connected to a 2900 metabolic cart (Sensormedics, Yorba Linda, CA). The patient achieved 2.39 L (53% predicted) and 3.09L (57% predicted) for FEV₁ and FVC, respectively. NHANES III Caucasian norms for ages 29 years and younger were used to determine predicted values (Hankinson et al., 1999). Reduced FEV₁, FVC, and FEV₁/FVC with a prior TLC of 4.65L (65% predicted), obtained with full-body plethysmography (6200 Autobox, Sensormedics Corporation) corroborate the patient's history of a mixed obstructive-restrictive ventilatory impairment. Baseline pulmonary function and plethysmography values appear in Table 1.

Table 1. Pulmonary function testing and plethysmography results. Data are absolute (percent predicted value) (NHANES III, Caucasian Norms, Hankinson et al., 1999).

FVC (L)	3.09 (57%)
FEV₁ (L)	2.39 (53%)
FEV₁/FVC	77
TLC (L)	4.65 (68%)
FRC(L)	4.30 (126%)
RV(L)	1.75 (123%)
D_LCO_{adj}	25.4 (68%)
MVV_{meas.}	125

Maximal breathing capacity was estimated by a maximal voluntary ventilation (MVV) test for 12 seconds at a ventilatory cadence of 90 breaths per minute. The patient achieved a breathing capacity of 125L, which exceeded the calculated MVV of 95.6L

that was derived based on FEV₁ multiplied by (Beck, 1997).

Cardiopulmonary exercise testing

Maximal exercise performance was measured using an incremental exercise test (IET) protocol performed on a cycle ergometer (Ergoline 800; Sensormedics, Corporation, Yorba Linda, CA) according to the guidelines of the American Thoracic Society/American College of Chest Physicians (2003) joint statement on cardiopulmonary exercise testing. The power output was continuously increased at in a step fashion at a rate of 20 Watts·minute⁻¹ to a symptom limited peak workload of 205 watts. The subject wore nose clips and breathed through a VMax Spectra mass flow sensor and Free Flow mouthpiece (Sensormedics, Corporation, Yorba Linda, CA). Expired fractional concentrations of oxygen and carbon dioxide were continuously monitored by a paramagnetic oxygen analyzer and non-dispersive infrared CO₂ analyzer (2900; Sensormedics, Corporation). Oxygen uptake (VO₂) and carbon dioxide output (VCO₂) were determined using standard algorithms. Breath by breath data were presented as a five breath rolling average. Resting measurements were made in the final 30 seconds of a three-minute stabilization period of breathing, after which the patient performed three reproducible inspiratory capacity (IC) maneuvers. The patient then performed unloaded cycling (zero Watts workload) for one minute followed by the step increase in power output. The patient was instructed to maintain a cycling cadence between 58-62 revolutions per minute. A 12 lead ECG (GE Case, Milwaukee, WI) was obtained at the end of each one-minute stage. Heart rate and peripheral oxygenation (Nellcor N595 Oximeter, Pleasanton, CA) were continuously recorded throughout exercise. Exercise tidal flow volume loops and IC maneuvers were repeated at two-minute intervals beginning at a power output of 40 Watts and were collected within 15 seconds of the termination of exercise. The stepped power output increased until the patient achieved volitional exhaustion. During the recovery period, the patient performed cycling at a power output of 20 Watts for a three-minute interval. Electrocardiographic monitoring was continued until the heart rate was near the observed resting rate.

The test was terminated due to leg fatigue with a Borg score of 10, and a dyspnea score of 10 reported at peak exercise. The patient exhibited excellent effort with a VO₂ ml·kg⁻¹·min⁻¹ that was 96% of the predicted value, and a respiratory exchange ratio (RER) of 1.2. A peak heart rate of 167 was attained which was 84% of the age adjusted predicted maximal value. A blunted blood pressure

Table 2. Maximal, symptom limited (exercise stop; leg fatigue-10, dyspne-10), incremental exercise tests (test increment; 20 Watts·min⁻¹) results.

	Peak	Predicted	% Predicted
Power (Watts)	205	200 †	(101%)
VO₂ (L·min⁻¹)	2.523	2.657 †	(96%)
VO₂ (ml·kg⁻¹·min⁻¹)	41.1	43.0 *	(96%)
ΔVO₂/ΔWR	11.5	8.7-11.9	
VT (VO₂ L·min⁻¹)	1.486	>1.062L >40% Pred. VO ₂	(56%)
HR (beats·min⁻¹)	167	200	(84%)
Heart Rate Reserve	33	<15bpm	
O₂ Pulse (ml·beat⁻¹)	15.11	13.29	(114%)
BP (mmHg)	152/94		
V_T (L)	1.7		(60%)
V_E/MVV_{meas}	123/125	~70%	(99%)
f (br·min⁻¹)	67	<60	
V_E/VCO₂ (at VT)	31	(N<34)	
RER	1.21	>1.12	

* American Heart Association (American College of Sports Medicine, 2000)

† Hansen Cycling Norms (Hansen and Wasserman, 1984)

response was observed with a peak blood pressure of 154/94. Left pleuritic chest pain rated as a 5/10 was reported which persisted until approximately 10 minutes post-exercise. No wheezing or dizziness was reported. CPET data appear in Table 2.

There was no significant reduction in aerobic capacity. Relative VO₂ was 41.1 mL·min⁻¹·kg⁻¹ (96% predicted), and absolute VO₂ was 2.523 L·min⁻¹ (96%) when based on American Heart Association (ACSM, 2000) and Hansen Cycling norms (Hansen et al., 1984), respectively. There was a normal ΔVO₂ to Δwork rate relationship (11.5). Presumably due to translocation of the heart, electrocardiogram indicated a right bundle branch block, determined to be not clinically significant. The patient otherwise demonstrated normal ECG and heart rate at rest and throughout exercise. The ventilatory threshold was normal: 1.486L (56% predicted), based on the dual criteria method for which the modified V-Slope and Ventilatory Equivalents method were used (Zeballos and Weisman, 1994).

Ventilatory responses revealed virtually total encroachment into ventilatory reserve as calculated by dividing the minute ventilation at peak exercise (V_E peak) of 123.3L by measured maximal voluntary ventilation (MVV_{meas}) of 125 L, a value of 0.99. Predicted V_E Peak/ MVV_{meas} is approximately 0.70 (Wasserman et al., 1999) which infers a significant

level of ventilatory constraint contributing to exercise limitation.

Ventilatory equivalents were normal at the ventilatory threshold (VT) as determined by the ventilatory equivalent for carbon dioxide (V_E/VCO₂). However, there was evidence to suggest hyperventilation near peak exercise based on ventilatory equivalents and ventilatory rate: V_E/VCO₂, V_E/VO₂, and ventilatory rate (Fb) were 45, 31, and 67 respectively. Graphically, extFVL provided evidence of ventilatory limitation and dynamic hyperinflation as there was clinically significant reduction in IC or greater than 200cc (470cc), and a corresponding increase in end expiratory lung volume (EELV). The data suggesting dynamic hyperinflation are reported in Table 3. Oxygen saturation did not substantially decrease from pre-exercise levels at the end of exercise (97% vs. 97%).

Forced spirometry was performed beginning at 5:00 post-exercise and continued at five-minute intervals until 20 minutes post-exercise, with no reduction in pulmonary function exceeding 7%. Consequently, there was no evidence of exercise-induced bronchospasm based on American Thoracic Society Guidelines for (ATS, 1991). Post-exercise spirometry data are reported in Table 4.

Table 3. Exercise tidal flow volume loop results (clinically, a drop of > 200cc in Inspiratory Capacity with a rising). End Expiratory Lung Volume is considered an indicator of dynamic hyperinflation.

	Baseline	40W	80W	120W	160W	205W
Inspiratory Capacity	2.37 L	2.38 L	2.46 L	2.62 L	2.64 L	2.15 L
End Expiratory Lung Volume	0.72 L	0.64 L	0.47 L	0.46 L	0.46 L	0.94 L

Table 4. Post-exercise (Post-ex) pulmonary function test results for exercise induced bronchoconstriction. Data are absolute (percent predicted value) (NHANES III, Caucasian norms, Hankinson et al., 1999).

	Baseline	Post-ex 5min	Post-ex 10min	Post-ex 15min	Post-ex 20min
FVC	3.09 (57%)	2.88 (-7%)	3.00 (-3%)	2.99 (-3%)	2.99 (-3%)
FEV ₁	2.39 (53%)	2.24 (-6%)	2.58 (8%)	2.43 (2%)	2.49 (4%)
FEV ₁ /FVC	77	78	86	81	83

TREATMENTS AND OUTCOMES

The patient was advised to maintain his current medication regimen for asthma, and to participate in daily physical activity to maintain conditioning level. The patient was accepted from the duty requirement for use of body armor.

DISCUSSION

With a significant body of literature that suggest exercise limitation in patients with unrepaired pectus excavatum is due to cardiovascular factors, the data in this particular case contradict conventional knowledge. The patient exhibited apparent ventilatory limitation, and terminated exercise with adequate heart rate reserve. In terms of the pre-test spirometry and patient's ability to achieve normal cardiovascular values for $\text{VO}_2 \text{ L}\cdot\text{min}^{-1}$, $\text{VO}_2 \text{ ml}\cdot\text{kg}^{-1}\cdot\text{min}^{-1}$, and $\text{O}_2 \text{ Pulse}$, the case is somewhat similar to the data reported by Castile et al. (1982). In their study, the mean total lung capacity was reduced and the test did not reveal alveolar-arterial oxygen difference abnormalities, effectively excluding significant ventilation-perfusion (V/Q) mismatching. In this case, the patient also exceeded predicted values for oxygen consumption, and V_E/VCO_2 at VT was normal at 31 and the absence of arterial desaturation argues against gas exchange abnormalities. Castile et al., suggest increased work of breathing may be responsible for the increase in oxygen uptake in such cases. This presumption follows logic, as during the inspiratory phase, the ventilatory musculature must overcome a non-complaint ribcage. This patient also suffered the disadvantage of dynamic hyperinflation that further exacerbated ventilatory mechanics. As a result, this case was not consistent with cardiovascular limitation as the source of exercise limitation. It also was an anomaly based on the report by Malek et al. (2003) that patients with a PSI of greater than 4.0 being eight times more likely to demonstrate a reduction in aerobic capacity compared to patients with a lower PSI, despite their level of exercise participation.

However, the ability of the patient to exceed predicted oxygen consumption may not be an unusual phenomenon in individuals greater than the age of 11 years. Patients with PE greater than this

age exhibit a tendency to "overachieve" whether academically, or athletically as a means to compensate for their deformity (Einseidel and Clausner, 1999). This particular patient appears to fit this profile as he was a competitive basketball player, and remains physically fit, and therefore was able to maintain "normal" functional capacity, despite the severity of his pectus deformity.

This patient showed clear clinical signs of ventilatory limitation demonstrated by a high V_E/MVV relationship, low tidal volume, and an abnormal ventilatory rate. It appeared the mechanical restriction and non-compliance of the chest cavity that was observed in the CT scan in figure 2, and was corroborated by 1) pulmonary function testing, 2) low tidal volume during exercise, and 3) the PSI, was adequate to restrict tidal volume expansion sufficiently to have caused dynamic hyperinflation and ventilatory limitation during exercise. Therefore, the patient's sole means to increase V_E is to increase the frequency of breathing. Despite a positive methacholine challenge test, the patient demonstrated a negative test for exercise induced bronchoconstriction which argues against ventilatory limitation due to bronchoconstriction.

CONCLUSIONS

Pectus excavatum has previously been associated with limitation of exercise (Beiser et al., 1972; Cahill et al., 1984; Peterson et al., 1985; Malek and Fonkalsrud, 2004). A large body of literature suggests that patients with pectus excavatum are most likely to have a cardiovascular limitation to exercise (Beiser et al., 1972; Bevegård et al., 1960; Bevegård, 1962; Garusi and D'Etorre, 1964; Haller et al., 1970; Shamberger, 2000) which is explained by mechanical restraint of the heart chambers and limitation of stroke volume.

Our case is novel in that our patient had a primary ventilatory limitation to exercise due to mechanical restriction of the chest cavity. Airflow obstruction from occult asthma was considered as a contributing factor to exercise limitation; however, post-exercise spirometry did not reveal bronchoconstriction. The patient demonstrated clear evidence of air trapping with increasing EELV during exercise and had an earlier positive confirmatory methacholine challenge test ($\text{PC}_{20} = 6.7$

mg·ml⁻¹). These findings can also be explained by bronchiolitis but there was no evidence for this seen on chest CT scan. The progressive air trapping in concert with chest wall restriction from his pectus excavatum satisfactorily explains the patient's exertional dyspnea. It is not surprising that the patient could not tolerate wearing a tight-fitting military protective vest due to breathlessness with even light exertion (e.g., walking). This would increase his chest wall restriction further, which would serve to oppose any increase in EELV. It is notable that the patient's relatively preserved VO₂ max despite these limitation points to his excellent effort and motivation.

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KEY POINTS

- Pectus excavatum (PE) is a relatively common phenomenon affecting approximately 1 in 300 births, with a 9:1 ratio of male to female rate of incidence.
- The etiology or exercise limitation is most frequently due to cardiovascular limitation due to the compression of the sternum upon the myocardium, impairing the ability to augment stroke volume.
- The Pectus Severity Index (PSI) is a useful indicator of pectus severity.
- Cardiopulmonary exercise testing provides useful data to distinguish between cardiovascular limitation, ventilatory limitation, or deconditioning in the evaluation of PE.
- In this case study, ventilatory limitation was due to the mechanical restriction of the thoracic cavity.

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