

Chest wall kinematics in young subjects with Pectus excavatum

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ABSTRACT

Quantifying chest wall kinematics and rib cage distortion during ventilatory effort in subjects with Pectus excavatum (PE) has yet to be defined. We studied 24 patients: 19 during maximal voluntary ventilation (MVV) and 5 during MVV and cycling exercise (CE). By optoelectronic plethysmography (OEP) we assessed operational volumes in upper rib cage, lower rib cage and abdomen. Ten age-matched healthy subjects served as controls. Patients exhibited mild restrictive lung defect. During MVV end-inspiratory and end-expiratory volumes of chest wall compartments increased progressively in controls, whereas most patients avoided dynamic hyperinflation by setting operational volumes at values lower than controls. Mild rib cage distortion was found in three patients at rest, but neither in patients nor in controls did MVV or CE consistently affect coordinated motion of the rib cage. Rib cage displacement was not correlated with a CT-scan severity index. Conclusions, mild rib cage distortion rarely occurs in PE patients with mild restrictive defect. OEP contributes to clinical evaluation of PE patients.

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1. Introduction

Pectus excavatum (PE) is the most common congenital chest wall deformity, resulting in a depression of the sternum and anterior chest. The size and shape of the depression range from mild concave depressions of a few millimeters to severe asymmetrical depressions of several centimeters. PE occurs in more than 1 of every 1000 births with a 3–4:1 male predominance and becomes more apparent during the period of rapid skeletal growth in early adolescence (Fonkalsrud, 1995).

Physicians and surgeons base the severity of PE on both clinical evaluation and CT scan (Haller et al., 1987). However, neither method measures the level of functional derangement of chest wall compartments in these patients. If the physiological impairments could be linked to the magnitude of the deformity and the latter could be adequately quantified, then it might be possible to identify those children at risk for complications in adult life (Wohl et al., 1995).

A mild reduction in lung volume (Derveaux et al., 1989; Morshuis et al., 1994) or presence of air trapping, even in patients with normal spirometry (Kelly et al., 2007; Koumbourlis and Stolar,

2004), have been reported in PE patients. Moreover it has been suggested that the depression of the sternum limits the movement of the ribs especially in the lower ones, thus preventing the expansion of the lower thoracic cross-sectional area (Koumbourlis, 2009). On theoretical grounds uncoordinated displacement of chest wall compartments is not unexpected in these patients, considering that a non-uniform distribution of pressure over the different parts may distort the rib cage (Chihara et al., 1996; Crawford et al., 1983; McCool et al., 1985; Sampson and De Troyer, 1982; Ward et al., 1992). However, recent studies (Aliverti et al., 1997; Kenyon et al., 1997; Romagnoli et al., 2006; Sanna et al., 1999) have shown that the expiratory action of the abdominal muscles plays a key role in minimizing rib cage distortion during exercise in healthy subjects. These data support previous observations showing that a normal swing in abdominal pressure with normal abdominal pressure–volume loop and normal tidal volume expansion are associated with normal rib cage mobility during increased ventilation in exercising PE patients (Mead et al., 1985).

It must be recalled that, usually, the maximal ventilation occurring during exercise is only about two-thirds of the maximal ventilation that can be achieved for brief periods of voluntary effort (Olafsson and Hyatt, 1969). Also, during maximal exercise test there can be a large negative pleural pressure during inspiration, but during expiration normal persons rarely, if ever, produce expiratory pleural pressures in excess of those that are required for maximal expiratory flow (Olafsson and Hyatt, 1969). This avoids the waste of muscular effort that would occur if expiratory flow became flow

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limited. In contrast, minimal pressure required to achieve maximal flow at a given lung volume is exceeded during maximal voluntary ventilation (MVV). When healthy subjects achieve MVV the pattern of ventilation is different from that achieved during exercise: tidal volume is smaller, respiratory frequency is higher, and end-expiratory-lung volume is greater than at resting functional residual capacity (FRC).

The question now arises. Should we expect a less coordinated activity of the respiratory muscles during MVV in PE patients? Preliminary results from our laboratory (Binazzi et al., 2009) indicating a normal reduction in end-expiratory volumes of the abdomen (suggestive of phasic expiratory abdominal muscle activity) and normal tidal volume expansion of both abdomen and chest wall prompted us to hypothesize that coordinated motion of the upper to lower rib cage prevents distortion during MVV in PE patients.

We evaluated chest wall kinematics during ventilatory effort in young patients with and without PE to validate the above hypothesis. Assessing volume displacement in each compartment allowed us to check the functional dimension of PE, the coupling of volume change between compartments and, if any, rib cage distortion, i.e., upper to lower rib cage uncoupling.

Accurate measurements cannot be obtained with methods unable to explore the tri-compartmental chest wall volumes during voluntary hyperventilation (Kenyon et al., 1997). Recently optoelectronic plethysmography (OEP) has been proven useful to assess volume changes of chest wall compartments, i.e., the upper rib cage (Vrc,p), lower rib cage (Vrc,a), and abdomen (Vab) during both spontaneous breathing and voluntary respiratory efforts (Aliverti et al., 1997; Binazzi et al., 2008; Kenyon et al., 1997; Lanini et al., 2003; Romagnoli et al., 2006; Sanna et al., 1999).

2. Materials and methods

2.1. Subjects

We studied 24 young male PE patients and 10 age-matched male controls. None of the patients had comorbidities. The study was approved by the local ethics committee and subjects gave their informed consent.

2.2. Protocol

All patients were well acquainted with the experimental protocol and equipment used. After baseline lung function measurements, optoelectronic measurements of chest wall motion were made with subjects sitting upright in a comfortable armchair. The measurements were made at rest during quiet breathing, which was defined as habitual comfortable breathing, and during MVV.

2.3. Pulmonary function tests

Routine spirometry, obtained with subjects seated in a comfortable armchair, was measured according to ATS/ERS guidelines (Miller et al., 2005). FRC was measured with a body plethysmograph (Autobox DL, 6200; SensorMedics; Yorba Linda, CA) according to a standardized procedure (Wanger et al., 2005). The normal values for lung function were those of Zapletal et al. (1972).

2.4. Maximal voluntary ventilation

12 s MVV was performed according to Miller et al. (2005). Tidal volume was set at 40% of vital capacity (VC), with a breathing frequency of ~90 breaths/min. Subjects wore a nose-clip and breathed through a low dead space mouthpiece. Flow was measured with a mass flow sensor (Vmax 229; SensorMedics) near the mouthpiece and lung volume changes were obtained by integrating the flow

signal. The flow rate at the mouth was recorded breath-by-breath. Cardiac frequency was continuously measured using a 12-lead electrocardiogram; oxygen saturation was measured using a pulse oxymeter (NPB 290; Nellcore Puritan Bennett, Pleasanton, CA, USA). The flow signal was synchronized to that of the motion analysis used for OEP and sent to a personal computer for subsequent analysis.

2.5. Additional experiments. Cardiopulmonary exercise testing (CPET)

In five patients symptom-limited cardiopulmonary exercise testing was conducted on an electronically braked cycle ergometer (Ergometrics 800, Sensor Medics, Yorba Linda, CA) using the Vmax 29c Cardiopulmonary Exercise Testing System (Sensor Medics). Subjects wore a nose-clip and breathed through a low dead space mouthpiece. Flow was measured with a mass flow sensor (Vmax 229; SensorMedics; 70 ml dead space) near the mouthpiece and lung volume changes were obtained by integrating the flow signal. After equipment was calibrated, each subject breathed at least 6 min through a unidirectional valve (Hans-Rudolph) while seated on the cycle ergometer with the expired air going into a universal exercise testing system (Ergometrics 800, Sensors Medics). Then they performed a 3-min warm-up of unloading pedalling followed by an incremental test. Patients were instructed to maintain the pedalling rate between 50 and 70 revolutions per minute. The patients were strongly encouraged to perform a maximal test, but they stopped volitionally exercise when they were no longer willing to tolerate the discomfort. A constant load cycle exercise testing under the anaerobic threshold (Vth) was performed 1 hour apart (for details see [Supplementary material](#)).

2.6. Optoelectronic measurements

OEP allows accurate 3-dimensional computation of the volume of the chest wall based on coordinates from surface markers attached to the chest wall surface. Details of this technique have been thoroughly described previously (Aliverti et al., 1997; Binazzi et al., 2008; Kenyon et al., 1997; Lanini et al., 2003; Romagnoli et al., 2006; Sanna et al., 1999).

2.7. Radiographic evaluation

Computed tomography (CT) of the chest was used to provide an objective assessment of the severity of the deformity. The severity was assessed on the basis of the Haller index which is the ratio of the transverse diameter of the thorax to the anteroposterior diameter at its narrowest point (Haller et al., 1987).

2.8. Analysis of the data

The OEP calculates absolute volumes, and the absolute volume of each compartment at FRC in control conditions was considered as the reference volume. Volumes are reported either as absolute values or as changes from the volume at FRC in control conditions.

The total chest wall volume (Vcw) was modeled as the sum of volume of the upper rib cage, i.e., the rib cage apposed to the lung (Vrc,p), volume of the lower rib cage, i.e., the rib cage apposed to the abdomen (Vrc,a) and volume of the abdomen (Vab). Thus, the Vcw was calculated as $V_{cw} = V_{rc} + V_{ab}$ and changes (Δ) in Vcw were calculated as $\Delta V_{cw} = \Delta V_{rc} + \Delta V_{ab}$. The time course of the volume of each region (Vrc,p, Vrc,a and Vab) along their sum (Vcw) was processed to obtain a breath-by-breath assessment of both ventilatory pattern and operational chest wall volume.

The presence of rib cage distortion was established by: (1) comparing the volume time courses in the upper rib cage (Vrc,p) vs.

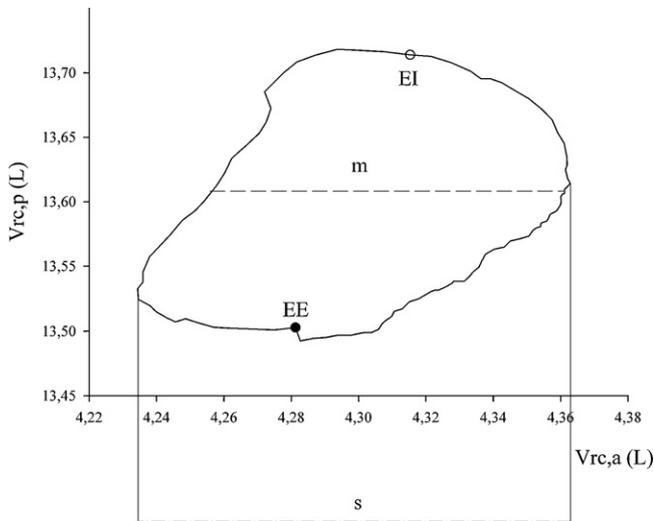


Fig. 1. Phase shift between Vrc,p and Vrc,a is indicated by the degree of opening of the Lissajou figure when the two volumes are plotted against each other. This was measured as the ratio of distance delimited by the intercepts of Vrc,p vs. Vrc,a dynamic loop on line parallel to the X-axis at 50% of RC,p tidal volume (m), divided by RC,a tidal volume (s), as $\theta = \sin^{-1} (m s^{-1})$. Vrc,p: upper rib cage volume; Vrc,a: lower rib cage volume; open circle: end-inspiration (EI); closed circle: end-expiration (EE).

volume in the lower rib cage (Vrc,a) and (2) by the phase shift between Vrc,a and Vrc,p when these two volumes were plotted again each other. This was measured as the ratio of distance delimited by the intercepts of Vrc,p vs. Vrc,a dynamic loop on a line parallel to the X-axis at 50% of RC,p tidal volume (m), divided by RC,a tidal volume (s), as $\theta = \sin^{-1} (m s^{-1})$, a previously adopted approach (Agostoni and Mognoni, 1966; Aliverti et al., 2009). In this system a phase angle of zero represents a completely synchronous movement of the compartments and 180° total asynchrony (Fig. 1).

The rest signals were recorded over a 3-min period after a 10-min period of adaptation to equipment. The volume tracings were

normalized with respect to time in each patient to allow ensemble averaging over the three reproducible consecutive breaths randomly chosen within the period of interest (at rest and during MVV and CPET at different times), and to derive an average respiratory cycle at each acquisition period. Inspiratory and expiratory phases of the breathing cycles were derived from the Vcw signal at zero flow.

Values are mean ± SD. Significance of changes in variables during maximal voluntary ventilation was evaluated by two-way analysis of the variance (ANOVA). Intra-group comparison was made by Wilcoxon and Mann–Whitney tests for paired and unpaired data with a 5% significance level. All statistical procedures were carried out using the PASW Statistics 18.0 (SPSS inc, Chicago IL, USA).

3. Results

Demographic characteristics were similar in the two groups, whereas lung volumes (FEV₁, VC and TLC) and MVV were significantly lower in PE group (Table 1).

3.1. MVV

3.1.1. Breathing pattern

The average tidal volume, respiratory frequency, driving and timing components of breathing pattern at rest (Fig. E2, Supplementary material and Table 2) were similar in patient and control groups.

Compartmental volumes at functional residual capacity, expressed as a percentage of chest wall volume, did not differ between the two groups (Table 3). Operational volumes of chest wall and its compartments at rest and during MVV are depicted in Fig. 2. End-inspiratory chest wall volume (Vcw,ei) ($p < 0.003$ and $p < 0.007$), end-inspiratory upper rib cage volume (Vrcp,ei) ($p < 0.0003$ and $p < 0.04$) and end-inspiratory lower rib cage volume (Vrca,ei) ($p < 0.004$ and $p < 0.0002$) all increased in PE and control groups. The end-expiratory chest wall volume (Vcw,ee)

Table 1
Anthropometric and lung function data in patients and controls.

	Sex	Age (year)	Height (m)	Weight (kg)	BMI (kg/m ²)	FEV ₁ (L)	FEV ₁ (%pred)	FEV ₁ /VC (%)	VC (% pred)	FRC (%pred)	TLC (%pred)	MVV (Lmin ⁻¹)	MVV (%pred)
Controls (10)	M												
Mean		14	1.71	67	23	4.27	110	85	107	106	110	121.4	88.4
SD		1	0.13	15	4	0.6	7	5	3	11	6	21.3	14.3
Patients (19)	M												
Mean		14	1.75	60	19	3.48	89	84	88	101	97	80.25	68.2
SD		2	0.12	14	3	0.81	14	7	15	19	14	15.3	22.9
<i>p</i>		0.24	0.96	0.39	0.26	0.06	0.01	0.71	0.01	0.49	0.049	0.01	0.01

BMI: body mass index; FEV₁: forced expiratory volume in one second; VC: vital capacity; FRC: functional residual capacity; TLC: total lung capacity; MVV: maximal voluntary ventilation.

Table 2
Breathing pattern in patients and controls.

	VT (%VC)	TI (s)	TE (s)	TI/TTOT	VT/TI (L s ⁻¹)	<i>f</i> (min ⁻¹)	VE (Lmin ⁻¹)
Controls							
Mean	18	1.11	1.24	0.47	0.72	26	20.4
SD	3	0.19	0.18	0.05	0.23	3	6.7
Patients							
Mean	18	1.24	1.52	0.46	0.57	24	15.5
SD	7	0.28	0.48	0.04	0.17	6	5.3
<i>p</i>	0.96	0.54	0.39	0.96	0.27	0.18	0.18

Values are means ± SD. VT: tidal volume; TI and TE: inspiratory and expiratory times; TI/TTOT: duty cycle; VT/TI: mean inspiratory flow; *f*: respiratory frequency; VE: minute ventilation.

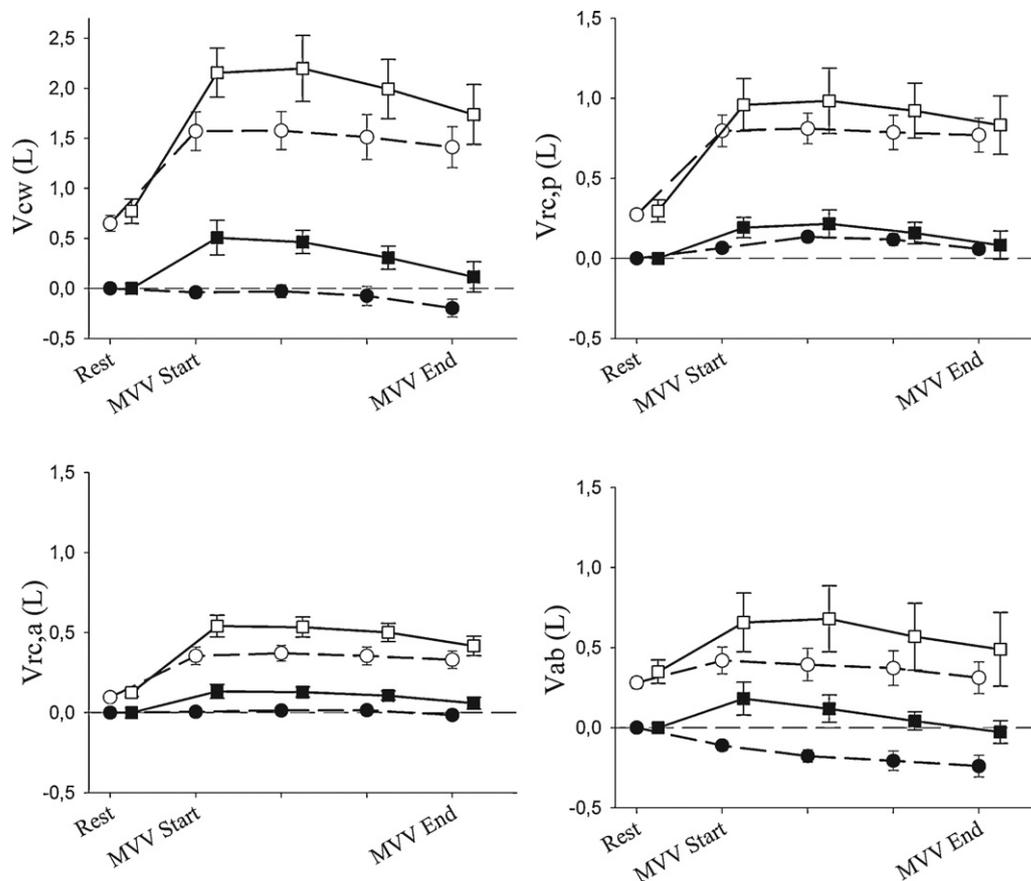


Fig. 2. Time course of the volumes of chest wall compartments from rest to end of maximal voluntary ventilation (MVV). Circles: patients; squares: controls; open symbols: end-inspiratory-volumes; closed symbols: end-expiratory-volumes. Vcw: chest wall volume; Vrc,p: upper rib cage volume; Vrc,a: lower rib cage volume; Vab: abdomen volume.

did not change ($p < 0.3$) in PE group because the decrease in end-expiratory abdomen volume (Vab,ee) ($p < 0.006$) was compensated for by an increase in end-expiratory upper rib cage volume (Vrcp,ee) ($p < 0.02$). In contrast, the end-expiratory chest wall volume (Vcw,ee) ($p < 0.04$), end-expiratory upper rib cage volume (Vrcp,ee) ($p < 0.04$), and end-expiratory lower rib cage volume (Vrc,a,ee) ($p < 0.05$) increased in control group. A comparison between the two groups at different MVV time intervals showed lower end-expiratory chest wall volume (Vcw,ee), end-expiratory lower rib cage volume (Vrc,a,ee) and end-expiratory abdominal volume (Vab,ee) ($p < 0.03$ for all comparisons) with a tendency for end-inspiratory chest wall volume (Vcw,ei) and end-inspiratory abdominal volume (Vrc,a,ei) ($p < 0.08$) to be lower in PE group. Between-group volume differences were no longer evident at the final MVV. In turn, PE and control groups kept similar constant tidal volumes of the chest wall (VTcw), upper rib cage (VTrc,p), lower rib cage (VTrc,a) and abdomen (VTab) during MVV, but PE patients set VTcw at lower operational volumes.

Table 3
Compartmental chest wall volumes at functional residual capacity.

	Controls	Patients	<i>p</i>
Vcw (L)	18 ± 5	16 ± 4	0.76
Vrc,p (% Vcw)	54 ± 3	54 ± 3	0.62
Vrc,a (% Vcw)	16 ± 1	14 ± 1	0.13
Vab (% Vcw)	30 ± 3	32 ± 2	0.88

Vcw: volume of the chest wall; Vrc,p: volume of the rib cage apposed to the lung; Vrc,a: volume of the rib cage apposed to the abdomen; Vab: volume of the abdomen.

3.1.2. Rib cage distortion

Individual data and group mean values of rib cage distortion measured by phase angle shift in PE and control groups during MVV are shown in Table 4. Values did not change significantly during the procedure in each group (ANOVA: $p = ns$). Using at least

Table 4
Phase angle degree in patients and controls.

	Patients			Controls		
	Rest	Start MVV	End MVV	Rest	Start MVV	End MVV
1	2.87	7.98	7.59	2.87	10.47	11.80
2	8.21	1.59	0.72	5.03	7.77	2.26
3	5.09	12.98	12.42	2.61	1.63	8.48
4	3.27	36.59	10.93	5.74	4.7	2.05
5	7.18	12.37	12.74	5.47	4.99	2.20
6	0	3.65	0.62	3.58	3.9	1.11
7	9.59	3.53	0	4.41	3.3	3.46
8	1.73	1.50	2.26	5.74	10.5	11.59
9	16.87	10.8	2.86	5.12	6.12	5.89
10	20.82	7.18	0	4.35	5.21	4.76
11	8.21	10.74	13.89			
12	6.34	10.48	11.54			
13	9.59	8.21	8.91			
14	4.59	5.22	8.85			
15	10.8	10.09	8.29			
16	15.46	2.55	2.66			
17	7.66	15.03	35.68			
18	5.21	25.37	28.73			
19	5.22	13.14	34.06			
Mean	7.83	10.47	10.67	4.49	5.86	5.36
SD	5.28	8.53	10.96	1.14	2.93	3.98

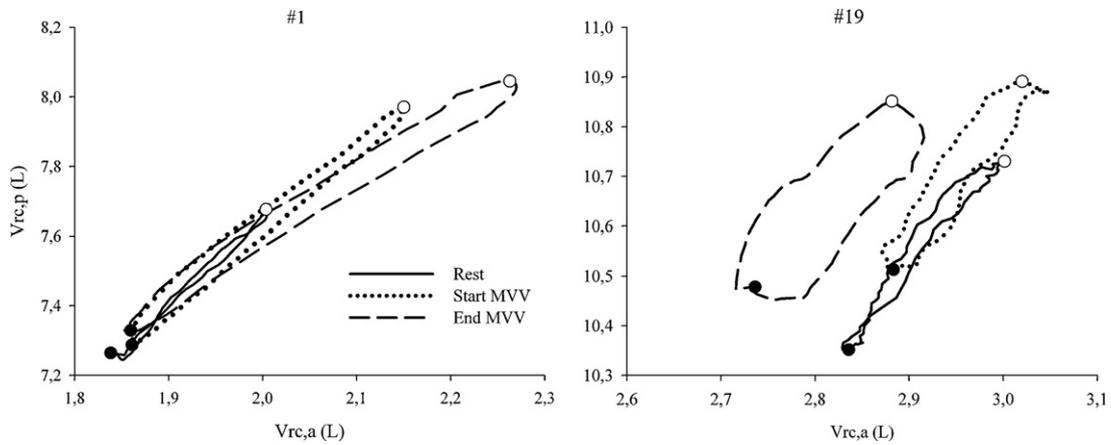


Fig. 3. Plots of $V_{rc,p}/V_{rc,a}$ in two patients without (left panel) and with (right panel) rib cage distortion from rest to maximal voluntary ventilation (MVV). Abbreviations as in Fig. 2; closed circle: end-expiration; open circle: end-inspiration.

2SD above the mean value for the normal subjects at rest and at MVV gave a threshold for the upper limit of normal of 6.8 and 13.3 phase angle degree, respectively. So mild rib cage distortion was evident at baseline in three patients (# 9, 10, 16), at start of MVV in two (# 4 and 18), and at the end of MVV in three (# 17–19). Plots of changes in upper rib cage volume ($V_{rc,p}$) to changes in lower rib cage volume ($V_{rc,a}$) in two representative patients (Fig. 3) show a phase angle of $<6^\circ$ in patients # 1 and # 19 at rest, but an early dynamic hyperinflation of the upper rib cage (increase in $V_{rcp,ee}$) with a phase angle shift of 34.06° during MVV in the latter patient.

3.2. Additional experiment: CPET vs. MVV

The five PE patients exhibited normal lung volumes at baseline. Operational volumes during MVV and CPET either increase or decreased (Δ from control) as follows: $V_{cw,ei}$ ($1.89L \pm 0.197$ vs. $1.42L \pm 0.25$, respectively; $p < 0.05$), $V_{rcp,ei}$ ($1.17L \pm 0.135$ vs. $0.78L \pm 0.125$, respectively; $p < 0.05$), $V_{rca,ei}$ ($0.428L \pm 0.078$ vs. $0.276L \pm 0.055$, respectively; $p < 0.05$), $V_{ab,ei}$ ($0.3L \pm 0.08$ vs. $0.37L \pm 0.20$, respectively; $p = ns$); $V_{cw,ee}$ ($0.06L \pm 0.058$ vs. $0.18L \pm 0.35$, respectively; $p = ns$), $V_{rcp,ee}$ ($0.316L \pm 0.16$ vs. $0.23L \pm 0.06$, respectively; $p = ns$), $V_{rca,ee}$ ($0.084L \pm 0.03$ vs. $0.10L \pm 0.06$; respectively, $p = ns$) and $V_{ab,ee}$ ($-0.36L \pm 0.13$ vs. $0.14L \pm 0.36$, respectively; $p = ns$). Fig. E3, Supplementary material depicts average changes in V_{cw} . No subject exhibited distortion

of the rib cage during MVV and exercise ($12.6^\circ \pm 2.6$, $4.9^\circ \pm 3.6$, respectively); two representative subjects are shown in Fig. 4.

3.3. CT-scan severity

A Haller index ≥ 6 was found in seven patients (# 1, 4, 6, 7, 10, 11, 16) with $<14^\circ$ phase angle shift at end MVV. No significant relationship was found between the Haller index and rib cage tidal volume during MVV ($V_{Trc_{mvv}}$) (Fig. 5), or any other kinematic variables.

4. Discussion

The novel findings of this paper are as follows: (i) early dynamic hyperinflation (increase in end-expiratory chest wall volume $V_{cw,ee}$) characterized chest wall kinematics during ventilatory efforts in young control subjects. In contrast, PE group set the boundaries of $V_{T_{cw}}$ at lower operational volumes (end-expiratory chest wall volume $V_{cw,ee}$ and end-inspiratory chest wall volume $V_{cw,ei}$) than control group. (ii) Mild rib cage distortion at rest was found in three patients, and only in another three patients did MVV impair the coupling of the upper to lower rib cage. (iii) The kinematic evaluation adds to the radiological evaluation of PE.

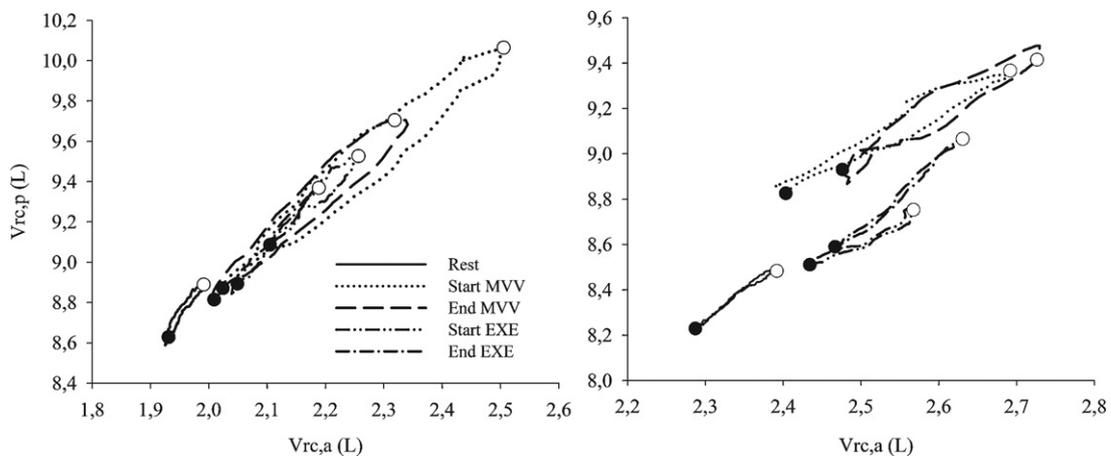


Fig. 4. Plots of $V_{rc,p}/V_{rc,a}$ in two representative patients with normal phase angle shift and mild dynamic hyperinflation (increase in $V_{rcp,ee}$ and $V_{rca,ee}$) during MVV and CPET.

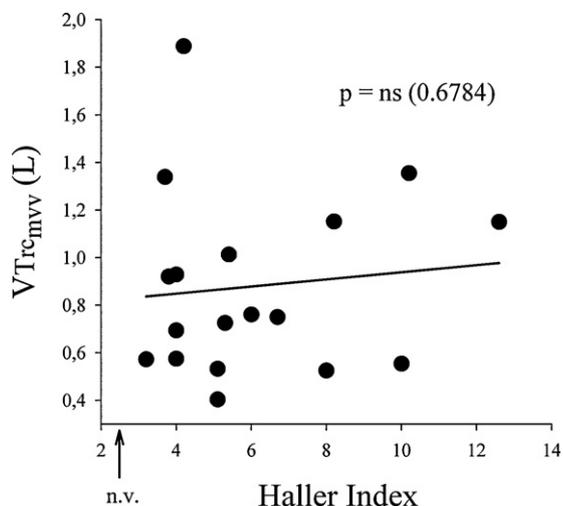


Fig. 5. Lack of relationship between Haller index and rib cage tidal volume during maximal voluntary ventilation ($V_{Trc_{mv}}$). Arrow indicates the normal value (2.5) of Haller index.

4.1. Critique of method

The limitations of optoelectronic plethysmography in assessing the relative changes in $V_{rc,p}$ and $V_{rc,a}$ have been thoroughly discussed in a previous paper of ours (Binazzi et al., 2008). Briefly, one limiting factor might be the changes in the cephalic margin of the zone of apposition, i.e., in the area over which the rib cage is effectively exposed to abdominal pressure (Chihara et al., 1996). However, the decrease in end-expiratory and a constant end-inspiratory abdominal volume during MVV are consistent with optimization of the diaphragm by preventing excessive shortening during inspiration (Aliverti et al., 1997; Kenyon et al., 1997; Romagnoli et al., 2006; Sanna et al., 1999).

4.2. Comments on results

Few detailed physiological studies have been carried out in young PE subjects either preoperatively or postoperatively (Mead et al., 1985). In the present study we first investigated chest wall kinematics at rest and during ventilatory effort with the aim of quantifying impairment and dimension of rib cage deformity in young people with and without PE. With OEP we provide a detailed description of volume displacement of the three chest wall compartments (Ward et al., 1992): the pulmonary rib cage (upper rib cage), abdominal rib cage (lower rib cage) and abdomen.

The pattern of chest wall kinematics found in healthy control group requires some comment. Dynamic hyperinflation is not unexpected in young healthy subjects during MVV when high expiratory pressure overcomes the minimal critical pressure required to achieve maximal flows at a given lung volume (Olafsson and Hyatt, 1969). In particular, dynamic hyperinflation of the chest wall (increase in $V_{cw,ee}$) was allocated in part in the lower rib cage ($V_{rc,a}$), promoting an increase in end-inspiratory volumes of the chest wall ($V_{cw,ei}$), and in part in the abdomen ($V_{ab,ee}$) to limit that increase.

A comparison of the data of the two groups shows that unlike control group, a decrease in abdominal volume ($V_{ab,ee}$) deflated the chest wall in PE group so that a constant tidal volume of chest wall (V_{Tcw}) was obtained at operational volumes lower than in control group. Our findings that PE and control groups kept similar constant tidal volumes of the chest wall, upper rib cage ($V_{Trc,p}$), lower rib cage ($V_{Trc,a}$) and abdomen (V_{Tab}) during MVV contrasts with the hypothesis that the depression of the sternum limits

the movement of the ribs (especially the lower ones) thus preventing the expansion of the lower thoracic cross-sectional area (Koumbourlis, 2009). The finding of lower end-inspiratory volume ($V_{cw,ei}$) and end-expiratory volume of the chest wall ($V_{cw,ee}$) in patients (Fig. E2, Supplementary material) indicates the need to keep $V_{cw,ei}$ far from the total lung capacity, with the aim of increasing inspiratory reserve volume and lower inspiratory muscle activation. On the other hand, the decrease in end-expiratory volume of the abdomen ($V_{ab,ee}$) promoting the decrease in end-expiratory volume of the chest wall ($V_{cw,ee}$), reflects an increase in expiratory muscle recruitment to deflate the abdomen (Aliverti et al., 1997; Romagnoli et al., 2006; Sanna et al., 1999).

Plots of upper rib cage volume ($V_{rc,p}$) vs. lower rib cage volume ($V_{rc,a}$) indicated a normal phase angle degree at rest and through MVV in control group. As stated by Crawford et al. "The maintenance of rib cage shape needs not be attributed to inherent stiffness but may be the consequence of apparently coordinated activity of the different respiratory muscles" (Crawford et al., 1983). Measurement of gastric and pleural pressure confirms that the effects of muscular activity on the shape of the rib cage simply reflect the combined influence of pleural and abdominal pressure (McCool et al., 1985). In PE patients, it has been suggested that the rib cage fails to move up and out during inspiration (Wohl et al., 1995). Available data, however, argue against this possibility. First, Mead et al. (1985) in a limited number of young patients, failed to demonstrate that PE is associated with decreased rib cage mobility. They reasoned that if rib cage expansion is limited, abdominal pressure should increase more during inspiration as was the case in young normal subjects during rib cage restriction. The lack of difference in abdominal pressure swings and pressure–volume loops between PE and control groups argues against decreased rib cage mobility in their patients. Also, lack of comparative measurements of lung and chest wall compliance is in contrast with the contention (Koumbourlis, 2009) that the decrease in chest wall compliance plays a role in abnormal rib cage motion in PE patients.

Due to the coordinated action of operating muscle forces, the pressure acting on the pulmonary and abdominal rib cage is nearly the same throughout the respiratory cycle. This results in very low rib cage distortion during leg exercise in healthy subjects (Duranti et al., 2004; Kenyon et al., 1997; Romagnoli et al., 2006). Our findings during both MVV and submaximal exercise make a case for a coordinated action of operating muscles underlying the undistorted rib cage configuration in the vast majority of PE subjects. Supporting our findings are data from Mead et al. (1985) showing that the increased slope of the volume/gastric pressure relationship, as PE patients increase exercise from rest to peak, reflects the greater contribution of the respiratory muscles other than the diaphragm which, when the slope changes sign, are the dominating influence. This pattern is of interest from the standpoint of respiratory muscle coordination.

4.3. Clinical implications of the study

The presentation of a patient with PE warrants a thorough workup to assess the significance of the defect. This workup can include those tests necessary to determine whether the patient should be referred to a surgeon for a discussion of repairs opportunity (Jaroszewski et al., 2010). The technique for quantifying the magnitude of PE defects has been developed based on measurements taken from chest X-ray, photographs of the body surface, and computed tomography. The most commonly used technique with reference values has been proposed by Haller et al. (1987). Discrepancies and controversies on the effects of corrective surgery in PE patients are at least in part attributable to the frequent lack of accurate evaluations of the severity of the deformation (e.g., by radiological indices) and even more by the lack of estimations of

relationships between functional variables and the degree of chest deformity. The results in the present study show the independence of methods for radiological and functional assessment of PE. In particular, patients with Haller index >6 did not exhibit rib cage distortion. This indicates the utility of investigating both methods in patients at rest, but also the need for an accurate evaluation of chest wall kinematics in conditions of increased ventilatory effort.

In conclusion, the assessment of chest wall kinematics helps quantify the physiology of rib cage deformity and adds to the radiological evaluation of patients with PE.

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Appendix A. Supplementary data

Supplementary data associated with this article can be found, in the online version, at doi:10.1016/j.resp.2011.11.008.

References

- Agostoni, E., Mognoni, P., 1966. Deformation of the chest wall during breathing efforts. *J. Appl. Physiol.* 21, 1827–1832.
- Aliverti, A., Cala, S.J., Duranti, R., Ferrigno, G., Kenyon, C.M., Pedotti, A., Scano, G., Sliwinski, P., Macklem, P.T., Yan, S., 1997. Human respiratory muscle action and control during exercise. *J. Appl. Physiol.* 83, 1256–1269.
- Aliverti, A., Quaranta, M., Chakrabarti, B., Albuquerque, A.L., Calverley, P.M., 2009. Paradoxical movement of the lower rib cage at rest and during exercise in COPD patients. *Eur. Respir. J.* 33, 49–60.
- Binazzi, B., Bianchi, R., Romagnoli, I., Lanini, B., Stendardi, L., Gigliotti, F., Scano, G., 2008. Chest wall kinematics and Hoover's sign. *Respir. Physiol. Neurobiol.* 160, 325–333.
- Binazzi, B., Coli, C., Innocenti Bruni, G., Gigliotti, F., Messineo, R., Lo Piccolo, R., Romagnoli, I., Lanini, B., Scano, G., 2009. Breathing pattern and chest wall kinematics in patients affected by pectus excavatum. *Eur. Respir. J.* 34 (Suppl 53).
- Chihara, K., Kenyon, C.M., Macklem, P.T., 1996. Human rib cage distortability. *J. Appl. Physiol.* 81, 437–447.
- Crawford, A.B.H., Dodd, D., Engel, L.A., 1983. Changes in the rib cage shape during quiet breathing, hyperventilation and single inspirations. *Respir. Physiol.* 54, 197–209.
- Derveaux, L., Clarysse, I., Ivanoff, I., Demedts, M., 1989. Preoperative and postoperative abnormalities in chest X-ray indices and in lung function in pectus deformities. *Chest* 95, 850–856.
- Duranti, R., Sanna, A., Romagnoli, I., Nerini, M., Gigliotti, F., Ambrosino, N., Scano, G., 2004. Walking modality affects respiratory muscle action and contribution to respiratory effort. *Pflugers Arch.* 448, 222–230.
- Fonkalsrud, E.W., 1995. Chest wall deformities. In: Rowe, M.I., O'Neill, J.A., Grosfeld, J.L., et al. (Eds.), *Essentials of Pediatric Surgery*. CV Mosby, St. Louis, pp. 383–391.
- Haller Jr., J.A., Kramer, S.S., Lietman, S.A., 1987. Use of CT scans in selection of patients for pectus excavatum surgery: a preliminary report. *J. Pediatr. Surg.* 22, 904–906.
- Jaroszowski, D., Notrica, D., McMahon, L., Steidley, D.E., Deschamps, C., 2010. Current management of pectus excavatum: a review and update of therapy and treatment recommendations. *J. Am. Board. Farm. Med.* 23 (2), 230–239.
- Kelly Jr., R.E., Shamberger, R.C., Mellins, R.B., Mitchell, K.K., Lawson, M.L., Oldham, K., Azizkhan, R.G., Hebra, A.V., Nuss, D., Goretsky, M.J., Sharp, R.J., Holcomb 3rd., G.W., Shim, W.K., Megison, S.M., Moss, R.L., Fecteau, A.H., Colombani, P.M., Bagley, T.C., Moskowitz, A.B., 2007. Prospective multicenter study of surgical correction of pectus excavatum: design, perioperative complications, pain, and baseline pulmonary function facilitated by internet-based data collection. *J. Am. Coll. Surg.* 205 (2), 205–216.
- Kenyon, C.M., Cala, S.J., Yan, S., Aliverti, A., Scano, G., Duranti, R., Pedotti, A., Macklem, P.T., 1997. Rib cage mechanics during quiet breathing and exercise in humans. *J. Appl. Physiol.* 83, 1242–1255.
- Koumbourlis, A.C., Stolar, C., 2004. Long growth and function in children and adolescents with idiopathic pectus excavatum. *Pediatr. Pulmonol.* 38, 339–343.
- Koumbourlis, C., 2009. Pectus excavatum: pathophysiology and clinical characteristics. *Paediatr. Respir. Rev.* 10, 3–6.
- Lanini, B., Bianchi, R., Romagnoli, I., Coli, C., Binazzi, B., Gigliotti, F., Pizzi, A., Grippo, A., Scano, G., 2003. Chest wall kinematics in patients with hemiplegia. *Am. J. Respir. Crit. Care Med.* 168, 109–113.
- McCool, F.D., Loring, S.H., Mead, J., 1985. Rib cage distortion during voluntary and involuntary breathing acts. *J. Appl. Physiol.* 58, 1703–1712.
- Mead, J., Sly, P., Le Souef, P., Hibbert, M., Phelan, P., 1985. Rib cage mobility in Pectus Excavatum. *Am. Rev. Respir. Dis.* 132, 1223–1228.
- Miller, M.R., Hankinson, J., Brusasco, V., Burgos, F., Casaburi, R., Coates, A., Crapo, R., Enright, P., van der Grinten, C.P.M., Gustafsson, P., Jensen, R., Johnson, D.C., Macintyre, N., McKay, R., Navajas, D., Pedersen, O.F., Pellegrino, R., Viegi, G., Wanger, J., 2005. Standardisation of spirometry. *Eur. Respir. J.* 26, 319–338.
- Morshuis, W., Folgering, H., Barentsz, J., van Lier, H., Lacquet, L., 1994. Pulmonary function before surgery for pectus excavatum and at long-term follow-up. *Chest* 105, 1646–1652.
- Olafsson, S., Hyatt, R.E., 1969. Ventilatory mechanisms and expiratory flow limitation during exercise in normal subjects. *J. Clin. Invest.* 48, 564–573.
- Romagnoli, I., Gorini, M., Gigliotti, F., Bianchi, R., Lanini, B., Grazzini, M., Stendardi, L., Scano, G., 2006. Chest wall kinematics and respiratory muscle action and dyspnea during arm vs. leg exercise in humans. *Acta Physiol.* 188, 63–73.
- Sampson, M.G., De Troyer, A., 1982. Role of intercostals muscle in the rib cage distortions produced by inspiratory loads. *J. Appl. Physiol.* 52, 517–523.
- Sanna, A., Bertoli, F., Misuri, G., Gigliotti, F., Randelli, I., Mancini, M., Duranti, R., Ambrosino, N., Scano, G., 1999. Chest wall kinematics and respiratory muscle action in walking healthy humans. *J. Appl. Physiol.* 87, 938–946.
- Wanger, J., Clausen, J.L., Coates, A., Pedersen, O.F., Brusasco, V., Burgos, F., Casaburi, R., Crapo, R., Enright, P., van der Grinten, C.P., Gustafsson, P., Hankinson, J., Jensen, R., Johnson, D., Macintyre, N., McKay, R., Miller, M.R., Navajas, D., Pellegrino, R., Viegi, G., 2005. Standardization of the measurement of lung volumes. *Eur. Respir. J.* 26, 511–522.
- Ward, M.E., Ward, J.W., Macklem, P.T., 1992. Analysis of human chest wall motion using a two compartment rib cage model. *J. Appl. Physiol.* 72, 1338–1347.
- Wohl, M.E., Stark, A., Stokes, D.C., 1995. Thoracic disorders in childhood. In: Roussos, C. (Ed.), *The Thorax Part C*, Marcel Dekker Inc., New York, pp. 2035–2069.
- Zapletal, A., Samanek, M., Tuma, S., Ruth, C., Paul, T., 1972. Assessment of airway function in children. *Bull. Physiopathol. Respir.* 8, 535–544.