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Correlating Haller Index and cardiopulmonary disease in pectus excavatum

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Abstract

BACKGROUND: The Haller Index (HI) has become standard for determining the severity of pectus excavatum. We compared patterns of cardiopulmonary dysfunction and their relationship with HI in patients with pectus excavatum.

METHODS: We performed cardiopulmonary exercise testing and chest computed tomography scans on 90 patients with pectus excavatum deformities at a regional pediatric hospital.

RESULTS: The median HI was 4.9 in patients with combined dysfunction, 4.4 in patients with isolated pulmonary dysfunction, 3.6 in patients with isolated cardiac dysfunction, and 3.4 in patients with normal function. HI varied significantly by disease group ($P < .009$). HI was significantly lower in patients with normal forced vital capacity than with abnormal forced vital capacity ($P = .001$). However, HI was similar in patients with normal and abnormal oxygen pulse ($P = .24$) or peak oxygen consumption ($P = .37$).

CONCLUSIONS: Fifty-nine percent of patients had cardiac and/or pulmonary limitation. A HI greater than 3.6 is associated with pulmonary dysfunction, but not cardiac dysfunction.

A major challenge in treating pectus excavatum (PE) is determining who would benefit from surgical repair, a decision that incorporates factors ranging from cardiopulmonary debilitation to the psychological impact of coping with a deformity. The Haller Index (HI), or pectus index, was proposed by Haller et al[1] in 1987 as an objective measure of pectus excavatum severity. HI is defined as the ratio of transverse to anteroposterior chest wall diameters, determined from a single axial computed tomography (CT) scan at the point of maximal pectus deformity.[1,2] In their study of 33 patients selected for surgical pectus repair and 19 age-matched control patients, Haller et al found that all the surgical cases but none of the controls had a HI exceeding 3.25, suggesting HI as an objective criterion for pectus excavatum repair.[1]

Anecdotal data suggest that pectus excavatum impairs exercise ability. However, patterns of cardiopulmonary dysfunction are not well delineated. Previous studies of preoperative pulmonary function showed a restrictive pattern that often is abnormal but rarely severe.[3,4] Patients with extreme

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HI are more likely to have pulmonary dysfunction manifesting as exercise intolerance, but a consistent relationship between HI and pulmonary function has not been shown across common HI ranges.[5] Postoperative lung function may either improve or worsen, depending on the repair and timing.[6,7] In terms of cardiovascular limitation, studies including meta-analyses disagree on the extent of preoperative cardiac dysfunction in pectus excavatum.[8,9] Despite findings of increased postoperative right ventricular and stroke volumes on echocardiography, both the clinical significance of the findings and correlation with HI are not well established.[10] We postulated that by stratifying patients by either pulmonary or cardiac disease, we might more clearly elucidate relationships between preoperative physiologic impairment and severity of chest wall deformity. We compared patterns of cardiopulmonary function, and their relationship with HI, in patients with pectus excavatum. Our hypothesis was that patients with either cardiac or pulmonary impairment would have a higher median HI.

Methods

We retrospectively reviewed the medical records of all patients who were referred to Seattle Children's Hospital, a regional, 250-bed pediatric hospital, for evaluation of pectus excavatum between January 2010 and May 2011. There were 115 patients who were potential surgical candidates and were referred for a preoperative assessment including the following: (1) cardiopulmonary exercise testing, including spirometry; (2) chest CT scan; and (3) surgeon evaluation. Patients who did not complete all 3 components were excluded.

Cardiopulmonary exercise test

Baseline spirometry and metabolic data were collected using the MedGraphics Ultima (Medical Graphics Corporation, St. Paul, MN) with BreezeSuite software. Exercise was performed on a cycle ergometer using a mouthpiece while metabolic data were collected. Three minutes of initial rest were followed by 3 minutes of unloaded pedaling at 60 rpm, then symptom-limited exercise, and 3 minutes of recovery at minimal workload. Heart rate, heart rhythm, and oxygen saturation were recorded continuously and blood pressures were recorded serially during the earlier-described period and through 10 minutes of recovery.

Pulmonary function

National Health and Nutrition Examination Survey III spirometry reference values appropriate for age, height, and sex were used to determine pulmonary function. Previous studies have varyingly defined pulmonary dysfunction as forced vital capacity (FVC) of less than 75% to 85% of predicted.[3,5] We defined dysfunction as less than 85% of predicted, and also performed a sensitivity analysis with a cut-off value of less than 80%.

Cardiac function

Test results were compared with predicted values for peak oxygen consumption and heart rate appropriate for age, height, and sex.[11,12] Patients who did not achieve a maximum heart rate of at least 70% the predicted value were deemed to have inadequate test results and were not included in the analysis. We defined cardiac dysfunction as either peak O_2 consumption (VO_2max) or oxygen pulse (equals O_2 consumption divided by heart rate) equal to less than 85% of predicted. Sensitivity analysis also was performed with a cut-off value of less than 80%.

Haller Index. Limited noncontrast chest CT scans were performed at our hospital on all patients, unless a previous chest CT scan of sufficient resolution was available, in which case CT scanning was not repeated. CT analysis and calculation of the Haller Index were performed by a board-certified pediatric radiologist (G.S.P.) in a blinded fashion from other patient data.

Statistical analysis

Characteristics of the patients included in the study were summarized descriptively overall and by cardiopulmonary disease group. HI then was compared across the 4 cardiopulmonary disease groups using the Kruskal–Wallis test. Separate post hoc Wilcoxon tests were performed to compare HI between patients with normal versus abnormal FVC, O_2 consumption (VO_2max), and O_2 pulse. Sensitivity analyses were conducted to assess robustness of the observed results to choice of cut-off value (80% compared with 85%) for each of the 3 cardiopulmonary measures. All analyses were performed using SAS (version 9.1.3; SAS Institute, Cary, NC).

Results

Ninety (78%) patients completed all study components; however, 3 patients were excluded from data analysis because of an inadequate maximal heart rate on the cardiopulmonary exercise test (CPET). Of 87 patients meeting inclusion criteria, 21 (24%) patients had isolated cardiac dysfunction, 16 (18%) patients had isolated pulmonary dysfunction, 14 (17%) patients had combined dysfunction, and 36 (41%) patients had neither (Table 1).

The median Haller Index was 3.7 (mean, 4.1; range, 2.3–11.3). The Haller Index varied significantly by disease group ($P = .01$), with the largest HI observed in patients with pulmonary dysfunction and with combined pulmonary and cardiac dysfunction (Fig. 1). Specifically, we found that HI was significantly higher in patients with reduced FVC

Table 1 Patient characteristics by disease group

	Overall (n = 87)	No disease (n = 36)	Pulmonary disease only (n = 16)	Cardiac disease only (n = 21)	Combined (cardiac and pulmonary) disease (n = 14)	*P* value
Male sex, %	80	94	75	95	100	.07
Mean age, y (SE)	15.2 (.3)	15.3 (.4)	15.4 (.7)	15.3 (.5)	14.7 (.5)	.84
Mean VO_2 max, % predicted (SE)	86.3 (1.7)	93.8 (1.6)	97.0 (1.9)	75.0 (1.5)	71.6 (2.0)	<.0001
Mean O_2 pulse, % predicted (SE)	98.4 (1.7)	104.7 (2.2)	112.9 (3.3)	85.6 (1.7)	85.0 (3.1)	<.0001
Mean FVC, % predicted (SE)	89.8 (1.4)	97.8 (1.5)	77.5 (1.5)	96.9 (1.8)	72.6 (1.8)	<.0001

than with normal FVC values (medians, 4.7 and 3.4, respectively; $P = .001$). However, HI was similar in patients with normal and abnormal O_2 pulse (medians, 3.5 and 4.0, respectively; $P = .24$) or VO_2max values (medians, 3.5 and 3.8, respectively; $P = .37$).

Most pectus excavatum patients show cardiopulmonary function less than 100% of predicted values, but the degree of each abnormality was modest in most cases. We used a scatterplot to compare continuous measures of pulmonary and cardiac function (FVC and VO_2max, respectively) (Fig. 2). By plotting patients according to degree of functional abnormalities, with dividing lines drawn at the 85% predicted point for each metric, the distribution of patients with each combination of functional limitation becomes apparent. In a quartile split based on HI, denoted by differential shading of the plotted points, patients with more severe HI (ie, >75th percentile) clearly cluster toward lower pulmonary function, not toward lower cardiac function.

We studied 2 metrics of cardiac function: VO_2max and O_2 pulse. The 2 metrics correlated linearly ($r = .88$; $P < .0001$). VO_2max consistently indicated a lower cardiac function than O_2 pulse as a percentage of predicted function, among patients both with and without cardiac limitation.

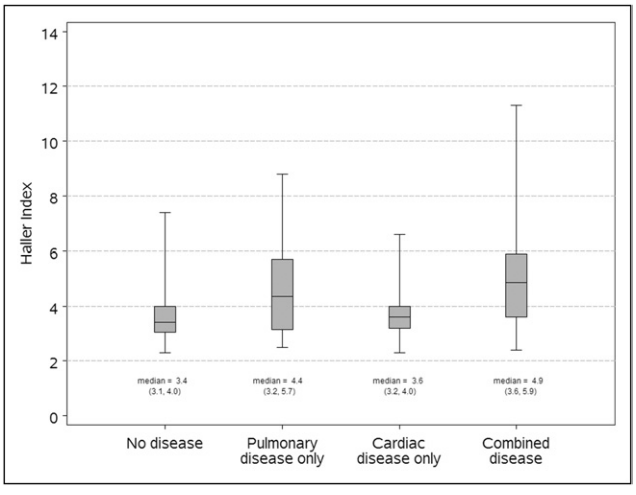

Figure 1 Haller Index differs based on pulmonary function: box plot of HI by disease group. Quartile values reported parenthetically for each disease group.

Similar results were observed in sensitivity analyses performed to assess the robustness of observed associations to definition of dysfunction (ie, <80% of predicted, rather than 85%). Based on this narrower definition of disease, 20% of patients had isolated cardiac dysfunction, 15% of patients had isolated pulmonary dysfunction, 10% of patients had combined dysfunction, and 55% of patients had neither.

Comments

We postulated that by grouping patients by pulmonary or cardiac limitation, we might elucidate relationships between the severity of chest wall deformity and physiologic impairment. We found that 59% of patients presenting for repair of pectus excavatum had cardiac and/or pulmonary dysfunction. A Haller Index greater than 3.6 was associated significantly with reduced lung function as measured by the vital capacity. However, an increased HI was not associated significantly with cardiac limitation. Patients with combined cardiac and pulmonary abnormalities had more severe cardiac and pulmonary impairment than those with an isolated abnormality, and had an even higher average Haller Index, averaging 4.9.

Advancements in surgical technique have made pectus excavatum repair safer and more widespread in the 25 years since the Haller Index came into use.[13] This proliferation has highlighted the importance of patient selection. The HI has gained popularity as a quantitative measure of PE, yet, a HI exceeding 3.25 originally was described as a fundamental feature of moderate or severe PE in patients selected for surgery based on an otherwise subjective evaluation. Haller et al[1] originally described the index as part of a preliminary report, their subsequent larger series did not correlate HI with any physiologic impairment.[14] Despite being a useful metric of deformity size, the clinical significance of the Haller Index remains unclear.

Patients with PE (or their parents) often report shortness of breath with exercise, but dyspnea at rest is rare. Presenting clinical concerns do not correlate with structural severity of the pectus deformity or with measures of cardiopulmonary limitation.[6,15] Given that most cardiopulmonary

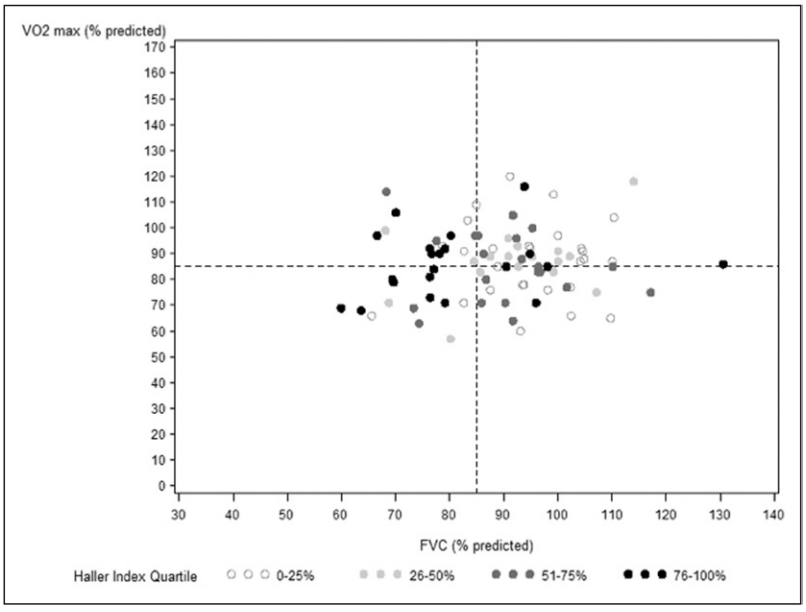

Figure 2 Distribution of Haller Index by cardiac and pulmonary dysfunction. Dotted reference lines at 85% predicted denote cut-off values for cardiac and pulmonary dysfunction, such that observations in the upper right quadrant show no cardiac or pulmonary dysfunction and observations in the lower left quadrant show combined (cardiac and pulmonary) dysfunction. Upper left and lower right quadrants represent isolated pulmonary and cardiac dysfunction, respectively. First quartile, HI of 2.3 to 3.1; second quartile, HI of 3.1 to 3.6; third quartile, HI of 3.6 to 4.7; and fourth quartile, HI of 4.7 to 11.3.

disease is subclinical, reliable methods for detecting and measuring cardiopulmonary dysfunction have proven elusive. Previous studies comparing pulmonary function with HI have shown discrepant results.[3,4,16] Studies comparing preoperative and postoperative spirometry generally show that pulmonary function worsens for several months after repair, but then returns to baseline and may improve after 1 year.[4,6] However, both of these studies reported patient-perceived improvement postoperatively, regardless of whether their spirometry measures improved. Lawson et al[5] argued that pectus excavatum historically has been treated as a binary phenomenon, "the patient either had it or did not have it," and proposed that it may better be seen as a spectrum of disease. They showed that at extremely severe HI values of 4 to 11, a correlation between HI and restrictive pulmonary disease emerges. Compared with the Lawson et al[5] study, most of our subjects exhibited a more moderate HI range of 3 to 7. We found that among patients with pulmonary dysfunction, median HI was more than 1 full unit higher, an average of 4.7 compared with 3.4 for patients without pulmonary disease.

The relationship between cardiac dysfunction and pectus excavatum is even more controversial.[8,9] Although exercise intolerance is associated with pectus excavatum, it often is thought to be multifactorial in nature with isolated cardiac dysfunction infrequently diagnosed on preoperative assessment. Although some researchers have found patients to have slight improvement in O_2 pulse and exercise tolerance after pectus repair, suggesting cardiac benefit, meta-analyses have questioned whether this improvement is statistically significant.[3,7] However, we identified cardiac limitation in 41% of patients with pectus excavatum. Nonetheless, it did not correlate consistently with the Haller Index. One hypothesis is that the right ventricle may experience compression earlier and with greater magnitude compared with the lungs, with resultant decreased right ventricular filling, because of its immediately substernal location and smaller relative volume. Alternatively, a pectus deformity may produce cardiac displacement rather than compression and thus have no impact on exercise tolerance.

We acknowledge several important limitations in this study. The retrospective, single-center design of this study was subject to inherent bias associated with subject selection. Because no consensus exists to distinguish normal from abnormal among CPET metrics, our use of 85% of predicted value is subject to scrutiny. However, our sensitivity analysis shows a similar distribution when lower cut-off value decrease inclusion into disease groups. Further, although we used a standardized exercise protocol for preoperative PE assessment, CPET is limited for assessing patients with substantially reduced exercise tolerance. However, 97% of subjects evaluated for this study achieved at least 70% of predicted heart rate. Finally, because we did not perform CPET studies postoperatively, we were unable to assess associations of preoperative HI severity with physiologic improvement after repair.

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Discussion

Dr Rana Ahmad (Spokane, WA): Patients with pectus excavatum mostly present with cosmetic complaints and rarely have significant physical limitations as perceived by them and their parents. An ideal index would be one which measures the functional disability in these patients and potentially predicts the improvement and outcome in their cardiac and pulmonary ability.

This study makes a good attempt at correlating the physiologic impairment with worsening Haller index. There is an increasing impairment in function with a rising Haller index. Patients develop both cardiac and pulmonary dysfunction over an index of 4.9. It is also worth mentioning that about 40% of the children had minimal physiologic dysfunction and underwent corrective surgery of their anatomic deformity. These patients may not see improvement on a functional level after surgery.

It would also be worthwhile expanding the study and assessing the improvement in cardiac and pulmonary function postoperatively. We may then be able to correlate the preoperative Haller index with the final outcome after surgery.