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The physiologic impact of pectus excavatum repair

Robert J. Obermeyer[a,b,*], Nina S. Cohen[b], Dawn E. Jaroszewski[c]

[a] Children's Hospital of The King's Daughters, Norfolk, VA, USA
[b] Eastern Virginia Medical School, Norfolk, VA, USA
[c] Mayo Clinic Arizona, Phoenix, AZ, USA

A R T I C L E I N F O

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A B S T R A C T

The adverse physiologic effects of pectus excavatum and subsequent resolution following correction have been a subject of controversy. There are numerous accounts of patients reporting subjective improvement in exercise tolerance after surgery, but studies showing clear and consistent objective data to corroborate this phenomenon physiologically have been elusive. This is partially due to a lack of consistent study methodologies but even more so due to a mere paucity of data. As experts in the repair of pectus excavatum, it is not uncommon for pediatric surgeons to operate on adult patients. For this reason, this review evaluates the contemporary literature to provide an understanding of the physiologic impact of repairing pectus excavatum on pediatric and adult patients separately.

Pediatric patients

Introduction

Pectus excavatum affects patients at different ages and is the most common congenital chest wall deformity (88%).[1] While some patients are born with the deformity, most do not develop the deformity until their prepubescent and early teenage years.[2] Preteen children are typically asymptomatic, but as they become more active in their teenage years they start to report symptoms. These symptoms include exercise intolerance, lack of endurance and shortness of breath with exercise.[3] Over 400 years ago in Spain, Bauhinus described a patient with severe pectus excavatum that suffered from exercise intolerance and since then there have been multiple similar reports in the literature.[4] More recent data supports this historic description with pectus patients reporting symptoms of shortness of breath and exercise intolerance ranging from 68 to 86%.[5,6] After repair, using validated questionnaires (i.e. Pectus Excavatum Evaluation Questionnaire (PEEQ), Child Health Questionnaire (CHQ)), a statistically significant perceived improvement in exercise tolerance has been reported by both patients and their parents.[5,7] A recent query of our database at Children's Hospital of The King's Daughters in Norfolk, Virginia from 1985 to 2018 which included 1270 patients demonstrated a similar finding. Approximately 95% of these patients no longer reported exercise intolerance with the bar in place. While these subjective reports are encouraging, it is important to investigate the anatomic and physiologic explanation for this perceived improvement after pectus excavatum repair.

In the previous decade there were several small studies that found evidence to support improved cardiovascular function after the Ravitch procedure.[8] The focus of this review is on data obtained from contemporary literature on pediatric pectus patients undergoing minimally invasive repair of pectus excavatum (MIRPE). The current literature frequently demonstrates improvements in pulmonary function,[9] chest wall mechanics,[10,11] cardiac function,[12] and cardiopulmonary exercise tolerance[13] after surgical correction of pectus excavatum. The dominant physiologic explanation is difficult to identify due to the complex interrelated function of respiratory mechanics and cardiac function, particularly during exercise. The current literature also has several limitations: short-term versus long-term results, rest versus exercise studies, inconsistent measure of pectus excavatum severity and outcomes, lack of control groups (i.e., normal patients or pectus patients forgoing treatment), inconsistent aerobic exercise capacity testing methods, and failure to control for conditioning of subjects. Finally, while a statistically significant change in exercise tolerance is difficult to demonstrate in even a well-designed study, the clinical improvement in exercise tolerance reported by the overwhelming majority of pectus patients after repair cannot be discounted.

Resting pulmonary function testing

The majority of pectus patients do not have pulmonary parenchymal or airway disease, therefore any improvement in pulmonary function after pectus excavatum correction is likely

* Corresponding author at: Children's Hospital of The King's Daughters, 601 Children's Lane, Norfolk, VA 23507, USA.

E-mail address: robert.obermeyer@chkd.org (R.J. Obermeyer).

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Table 1
Percent predicted FVC before repair and after bar removal.*

Author	Year	Patients	Pre-operative	Post bar removal	*p-value*
Lawson**	2005	25	84 ± 8	90 ± 11	**0.0015**
Castellani	2010	46	91 ± 14	88 ± 13	0.117
O'Keefe	2013	67	91 ± 19	99 ± 23	**<0.001**
Maagaard	2013	44	92 ± 14	92 ± 13	n.s.

* Values rounded to whole numbers for clarification and ranges estimated based on available data.
** Includes only patients over 11 years of age.

Table 2
Percent predicted FEV1 before repair and after bar removal.*

Author	Year	Patients	Pre-operative	Post bar removal	*p-value*
Lawson	2005	25	81 ± 8	90 ± 9	**0.0052**
O'Keefe	2013	67	81 ± 17	90 ± 21	**<0.001**
Maagaard	2013	44	86 ± 13	93 ± 13	**<0.001**

* Values rounded to whole numbers for clarification and ranges estimated based on available data.

attributable to an improvement in respiratory mechanics.[10] At rest the diaphragm initiates most of the inspiratory effort. During exercise, more thoracic excursion is required to generate higher lung volumes, at which time the sternum and costochondral cartilage become more important for efficient respiration.[14] Although resting pulmonary function tests (PFTs) may not be the optimal way to evaluate patients complaining of symptoms during exercise, they have the inherent strength of being reported against a normal distribution.[15]

Siglet et al. from Calgary produced one of the first comprehensive reports in 2003 which looked at results 3 months after repair in 11 patients.[16] This investigation helped pave the way for future studies by including an assessment of exercise tolerance, pulmonary effects, and cardiac function. Their patients universally reported modest to marked improvement in exercise tolerance, but interestingly their PFTs worsened. This result was likely secondary to patient deconditioning as reported by the authors. One month later, Borowitz and the group from Buffalo performed similar testing on 10 patients, for whom the deleterious effects on PFTs resolved at 6 –12 months after repair.[17] Many authors have even made the argument that PFTs may be suboptimal while the bar is still in place due to its restrictive effects. For this reason, the literature reviewed here includes only long-term studies where PFTs were compared before repair and after bar removal. The first study to compare PFT outcomes at these two time points was reported by Lawson et al. in 2005. This study included 45 patients with an FVC and FEV1 measured anywhere from 0.1 to 3.8 years (average 1.2) after bar removal. Patients less than 11 years of age did not appreciate a significant increase in PFTs, however those over 11 years of age had a 6% increase in FVC and a 9% increase in FEV1, both reaching statistical significance.[18] Five years later, Castellani et al. completed a cardiovascular performance study in Austria which was similar to Sigalet's study in 2003. Only the long term outcome of FVC was reported, which decreased but did not reach statistical significance.[21] Almost a decade after his first report, Sigalet collaborated with O'Keefe et al. to publish the follow-up study "Longer term effects".[12] This study also detailed the effects of MIRPE on PFTs and demonstrated a significant improvement in both FVC and FEV1 after bar removal.[12] In the same year, Maagaard et al. found no significant change in FVC but a statistically significant increase in FEV1 after MIRPE[13] (Tables 1 and 2). Maagaard et al. was the only group to examine PFTs in pectus patients versus matched controls. A statistically lower FEV1 in preoperative pectus patients (48) versus controls (25) (86% vs 94%; p=0.008) was noted, but this normalized 3 years later after pectus bar removal (93% vs 97%; p=0.268). There was no statistically significant change in FVC before repair (92% vs 98%; p=0.060) or 3 years later (92% vs 98%; p=0.076).[13]

It is worth noting that changes in PFTs after the Ravitch procedure have been less favorable than after MIRPE, potentially due to the disruption and subsequent calcification of the costochondral cartilage.[20,21] This concept combined with the greater improvement appreciated in FEV1 over FVC in all of these studies (Tables 1 and 2) may help provide some explanation for improved exercise tolerance. One theory to explain this is that the total volume of air that can be exchanged during maximal inspiration and expiration (i.e., FVC) is not significantly affected by predominantly reshaping the middle of the chest wall. However, the remodeling of the costochondral cartilage by MIRPE may improve respiratory mechanics such that patients are able to expel air faster in one second (i.e., FEV1) and thus inhale oxygen-rich air faster during exercise. In summary, resting PFT studies evaluating FVC and FEV1 show an expected initial decline shortly after MIRPE that improves to slightly above baseline after bar removal in most studies.

Chest wall motion analysis

In 2011, Redlinger et al. employed Optoelectronic Plethysmography (OEP), a form of motion analysis, to demonstrate regional chest and abdominal wall motion dysfunction in pectus excavatum patients. During deep breathing, the movement of the upper and lower sternum was decreased by 28–51%, and the abdominal wall motion was increased by 147% in pectus patients compared to matched controls. The significant increase in abdominal wall motion was hypothesized to be a compensatory reaction to a relatively fixed sternum during forceful breathing.[10] The following year, 42 patients underwent repeat testing 6 months postoperatively with the Nuss bar in place. There was a resolution of aberrant paradoxical movement of the sternum and abdominal wall during forceful respiration. These studies demonstrate an increase in sternal motion after pectus excavatum correction, which may provide a biomechanical explanation for the noted increase in FEV1. The improvement in respiratory mechanics as demonstrated by OEP analysis and PFT studies may help explain patient reported improvement in exercise tolerance after MIRPE.[12,22]

Resting cardiac function testing

Right ventricle

Analogous to resting PFTs, cardiac studies performed during rest may not provide a clear physiologic cardiac explanation for improved exercise tolerance. Jeong et al. demonstrated a statistically significant resolution of cardiac compression, namely the right ventricle, on computed tomography after pectus excavatum repair. However, it is important to recognize that supine imaging may underestimate the severity of this compression when the patient is in the upright position during exercise.[11] This cardiac compression was further characterized by Coln et al. in 2006. They reported the largest pediatric series of preoperative and postoperative resting echocardiograms in 123 pediatric patients with pectus excavatum and demonstrated that 117 pectus patients had resolution of chamber compression after Nuss repair. Valve abnormalities, most commonly mitral valve prolapse and regurgitation, also resolved in almost all patients.[6] These anatomic changes produce a relief of cardiac compression that has been postulated by several authors to improve right heart filling and function.[6,11,23] In 2016, Topper et al. proved this effect using Cardiovascular Magnetic Resonance (CMR) imaging. CMR has evolved to be the accepted standard to evaluate the right ventricular function due to its complex crescent shape that makes it difficult to model with