Long-term results following surgical correction of symptomatic pectus excavatum in seniors. Considerable improvement of cardiovascular function and quality of life.

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Introduction

Pectus Excavatum (PE), also known as funnel chest, is a diagnosis usually made at a young age. It is a congenital defect, distinguishable by a funnel-shaped dent in the medial anterior thoracic wall at the level of the 4th -7th rib.[1,2] In Europe, this abnormal chest shape is found in about 1:1000 births with the majority of cases being male.[3] There are syndromes such as Ehlers Danlos, Marfan and Poland, where correlation with PE is mentioned.[1-4] Experts are unsure of the exact cause, although often there is a familial/genetic predisposition.[5-7] Children and adolescents with this diagnosis may have symptoms that are cosmetic or psychological in nature. However, there are cases of severe respiratory problems and difficulty when exercising. These symptoms occur as a result of cardiac compression, due to the sternum exerting pressure on both the right atrium and right ventricle.[1,8-14] This also explains the "postural component": respiratory distress and palpitations increase by bowing to the front and decrease in overstretching the back together with pulling both arms backward[15,16]. Less well known, is that this defect may also result in serious problems later in life. This is caused by the fact that the rib cartilage loses its flexibility at an older age.[10,15,17] This leads to the heart becoming less able to rotate, causing, in particular, problems with the right atrium. We refer to this as Symptomatic Pectus Excavatum in Seniors (SPES).[16] Until 20 years ago, this diagnosis was not recognized as a serious problem by general practitioners and specialists[1,10,14,18] PE was seen more of a cosmetic issue than a deformity of the chest that could cause serious problems. Subsequently, there have been many more publications significantly raising awareness amongst specialists.[1,2,3,14,19-23] There have been a number of small studies, two of which were from our group, focusing on the short term outcomes of surgical repair in seniors (> 40years) with PE.[15,16] These earlier studies revealed a beneficial effect on cardiovascular function and QoL. However, we were unable to find any publications that considered the long term outcomes of surgical repair in these patients. This is one of the first follow up studies, where a significant number of surgical patients (n=30) are followed for a longer period of time (mean 4.4 years).

Materials And Methods

This study was undertaken by a team of specialists from the Surgical and Cardiology departments of Zuyderland MC Heerlen. The research study was approved by the Local Research Ethics Committee in April 2014 and informed consent was obtained from all participants involved. Patients were interviewed by phone or seen in our outpatient department between May and September 2014.

Inclusion criteria

Our research included patients 40 years of age and older with SPES who had undergone reconstructive surgery in the period from 2007 to 2014. Excluded were patients from our database who were not able to attend the outpatient department and could not be interviewed by phone. Compared to previous research, we lowered the age limit of SPES from 50 to 40.

This was done in consultation with the surgeon and based upon the fact that no difference was found in elasticity of the sternum at age 40 or 50.

Design

The information obtained from the group of patients attending the outpatient department included medical history, physical examination, electrocardiography, transthoracic echocardiography, medical photography, spirometry, X-Ray thorax and an MRI or CT heart scan.(Figures 1a,b and 2a,b). For patients where only interviewing by phone was possible, only medical history was documented. By using this information we were able to evaluate patients' complaints over the last year.

To assess if and how patients continued to suffer from symptoms related to SPES, new signs, symptoms and eventual test results, were arranged in a SPES score.[16] This score ranges from 1 to 10 giving each finding a severity score (Table 1). This score gives information about the patient's symptoms. A score of 0, indicates that the complaints and symptoms are not attributable to the PE. Scores of 5 or more indicate that the symptoms are attributable to an existing pectus excavatum(PE). The SPES score differs from the one used in previous studies. The adjustments made were based on the findings in a previous article by Kragten et al. that a Treadmill ECG and expiratory lung function tests had no significant value[16]. We therefore chose to measure only inspiratory restriction using a Triflow Incentive Spirometer and tested patients with and without postural provocation. This contrasts with other published research where expiratory restriction was measured with varying and mostly unconvincing results.[1,24,25,30-33] To assess the cardiac indentation and pump function we found that an MRI scan post operatively was the preferred procedure. To achieve the correct evaluation, when performing an MRI, it is essential that the patient provokes the postural symptoms by bending the head downwards. In our appraisal of previous studies, we have noted that this was not always the case. The patients thoracic X- Rays and MRI scans were reviewed by a dedicated radiologist. In our previous research, we found that were of no value[16]. In addition to the SPES score, we also used a specific PE questionnaire. Subjective QoL improvement was assessed by asking patients the question: 'How was your quality of life before and after the operation expressed as a number on a scale of 1 to 10 ?'. Beforehand patients were instructed about the QoL in relation to exercise capacity and overall wellbeing. Finally, medical photographs were taken to document longer-term cosmetic results[34].

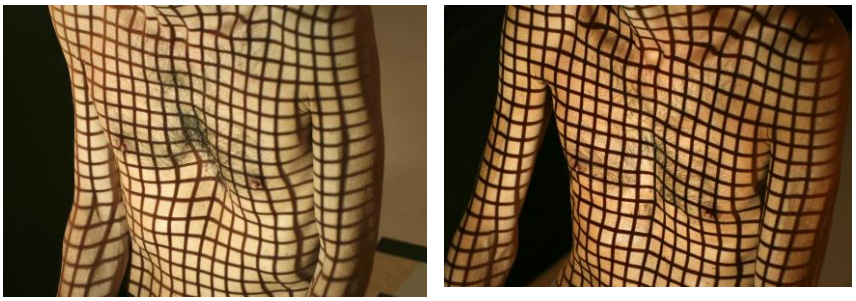


Fig 1
Pre and postoperative 3D grid projection photograph

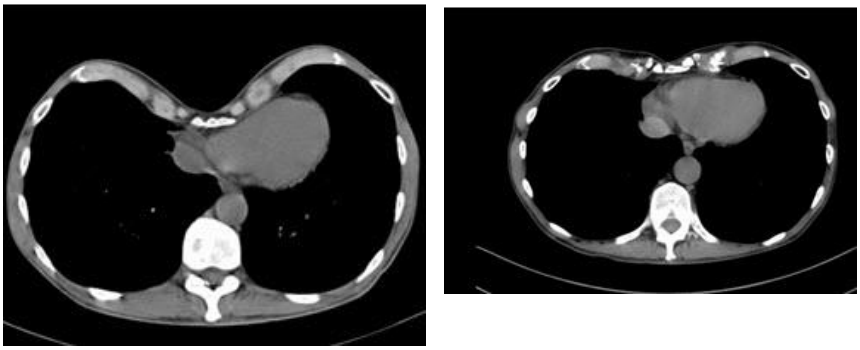


Fig 2
Pre- and postoperative CT

Clinical Finding		0	1	2	3
Dyspnoea		No	Minor	Moderate	Severe
Palpitations		No	Minor	Severe	
Fatigue, low exercise tolerance		No	Minor/Moderate	Severe	
Chest Pain		No	Yes		
Postural Symptoms		No	Possible	Clearly Present	
Arrhythmia	(ecg)	No	Clearly present	Severe	
Mitral valve insufficiency	(echo)	No	Minor	Clearly Present	
Cardio compression	(MRI/CT)	No	Yes		
Inspiratory obstruction	(Triflow)	No	Moderate	Clearly Present	

Table I **Spes Scores**

Postoperative patients were monitored by the surgeon and/or cardiologist over a period of 12 months. Following this period, patients were not seen by a specialist and were subsequently seen by a doctor only if there were complications. For this reason the authors wished to evaluate these patients after a minimum post-operative period of one year.

Data collection and Statistical analysis

The medical data for these patients was collected from two sources. One source was a dedicated MS Access-2001 database, specially developed for similar research in the past16. The second resource was the general hospital database MEDVIEW. These databases were used for analysis, compiling reports and collection of new patients. All analyses were carried out with SPSS statistics version 20.0 and Microsoft Excel 2010. Data was presented as means +/- SD or percentage. In addition, we compared the baseline values prior to our publication in 2011 (immediately preoperative), with those in 2014. P-values less than 0.01 were considered as statistically significant.

Results

34 patients over the age of 40 had surgical correction of their PE using either the Ravitch or NUSS bar procedure. [1,10,13,22] These patients were closely supervised and monitored by the interdepartmental team. Of this group, 4 patients elected not to participate in our group. This gave a total of 30 patients. Of these patients, 15 were unable to attend the outpatient department. The reasons given were; travelling distance, counting up to over 250 KM, financial issues and lack of interest. They agreed to complete our questionnaire either by telephone or email. The group comprised patients in the age category of 44 to 77 years undergoing surgery between August 2007 and May 2013.

General characteristics	Targets	Conservatives
Age (years)	57,9 +/- 10,4	62,4 +/- 10.0
Sexe (%)		
men	73,3%	72,5%
women	26,7%	17,5%
Genetic load	60,0%	31,7 %
Pulmonary Disease	10,0%	21,9 %
Coronairy Artery Disease	10,0 %	12,2 %
Smoking Status	13,3 %	21,9%

Table II Clinical characteristics of the study population at follow up

Most frequently cited complaints were; dyspnea and postural symptoms. The group was divided into two subgroups; one for patients aged 50 years or older, the other for patients aged 40 - 49 years. A modified Ravitch procedure was used in patients from the senior subgroup and the NUSS Bar procedure for patients aged 40 – 49 years. General characteristics of the patients are shown in Table 2.

Clinical outcomes

Following surgery, most patients experienced a considerable improvement and reported either a few or no significant symptoms. On questioning, the remaining symptoms experienced by a number of patients included shortness of breath, tiredness or palpitations (Table 3a,b).

In terms of side effects one patient described a cough reflex and postural shortness of breath. This was caused by pseudo-arthrosis of the sternum 7 years post-surgery. Other adverse events in the immediate post-operative period were bleeding and infection (n=3). Most patients were satisfied with the cosmetic result, 9 patients were not satisfied with the cosmetic outcome of their operation. Analyzing the results we compared the symptoms and clinical signs of the patients prior to surgery before our first publication in 2011 and those of these patients in 2014 (Figure 3). We saw a significant and steady postoperative decrease in postural symptoms (p<.001) and there was a significant difference in dyspnea (p<.001), palpitations (p<.001)) and QoL (p<.001) when comparing 2011 and 2014 (Table 3a,b).

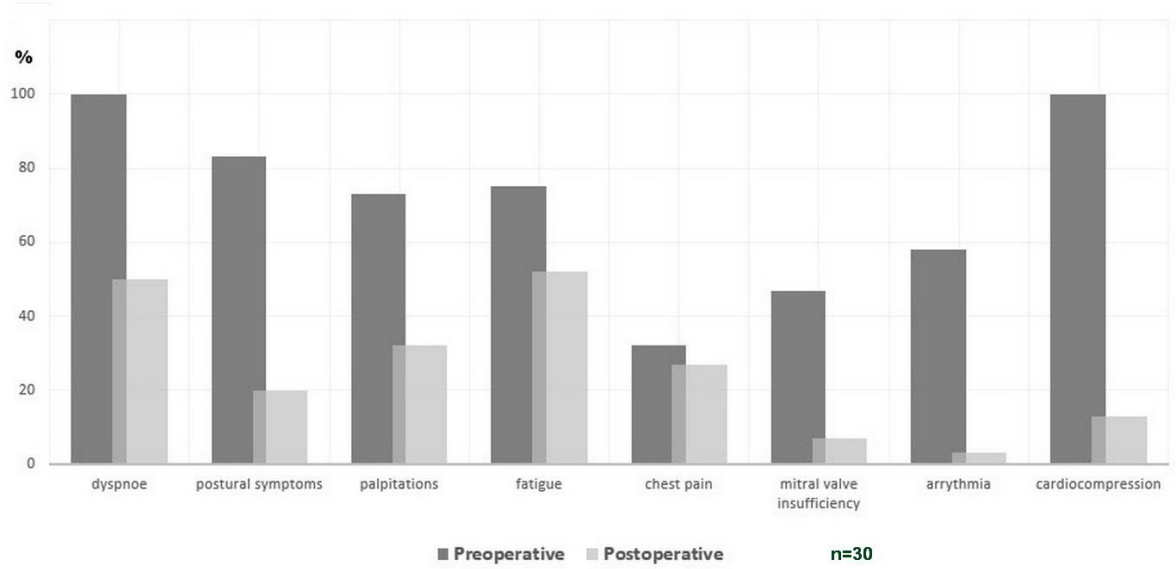
Preoperative Clinical Finding	**0**	**1**	**2**	**3**
Dyspnea	0	5	19	6
Palpitations	5	13	12	
Fatigue, low exercise tolerance	8	13	9	
Chest Pain	22	8		
Postural Symptoms	0	5	25	
Irregular ECG tracings	14	3	13	
Valve insufficiency on echocardiogram	16	9	5	
Cardiocompression on MRI/CT	0	30		

Table IIIa SPES scores Targets preoperative

Postoperative Clinical Finding	**0**	**1**	**2**	**3**
Dyspnea	15	10	4	1
Palpitations	20	15	1	
Fatigue, low exercise tolerance	22	8		
Chest Pain	22	8		
Postural Symptoms	24	4	2	
Irregular ECG tracings	15			
Valve insufficiency on echocardiogram	14	1		
Cardiocompression on MRI/CT	11	4		
Spirometry (Triflow)	14	1		

Table IIIb SPES scores targets postoperative

There were however no significant differences in fatigue and chest pain. There was no postoperative improvement in 20% of the 30 patients (n = 6), they retained a SPES score ≥ 5, they continued to suffer from SPES and we saw only minimal cosmetic improvement on their medical photographs. Mitral valve prolapse was seen in the transthoracic echogram of one of these patients, heart compression was still present in 4 patients and one patient experienced difficulties in performing the Triflow test.



The clinical results of surgical correction of Pectus Excavatum were good to excellent in most cases. Symptoms present prior to surgery, such as dyspnea, postural symptoms, fatigue and palpitations, were substantially reduced or had disappeared completely.

Discussion

The aim of this study was to investigate, over the long term, the treatment and clinical outcomes of patients with SPES. These patients were referred after a follow-up period of at least one year.

The main finding of this study is that symptoms of shortness of breath, palpitations, reduced exercise tolerance and overall QoL issues continued to be significantly improved after surgery, when compared with the results of our earlier studies.[15,16] In recent years there has been increasing interest in PE patients. The results of our follow up study of patients with SPES are in accordance with earlier research performed on the same group. Our investigations indicated that chest radiography should be used to evaluate any side effects of surgery. To provide additional information on cardiac compression an MRI with postural provocation was required to determine if the symptoms were caused by the presence of PE. Cardiac compression was still evident on the MRI of four patients. This may be explained by the fact that these patients had the deepest indentation. As a result, surgery provided relief of symptoms, but did not resolve the cardiac compression. The severity of the defect could thus be correlated with the final surgical outcome. In comparison to our earlier research, the new SPES score gives a more detailed picture of the patient's complaints over the longer term. This means it can be used for further purposes in the future. Measuring inflow restriction with an incentive spirometer during postural provocation provides very specific data in some SPES patients. Therefore, it should be routinely performed in all patients. Based upon literature research, we adopted the trans-thoracic echocardiogram (TTE) as a reliable tool to evaluate cardiac functions after surgery. Dawn et al. evaluated a patient with PE by measuring the degree of RV compression. They concluded that, when correctly performed, a TTE can accurately provide clinically relevant information about cardiac size and any hemodynamic compromise in patients with PE.[35] By contrast, our study showed it was

difficult to obtain information on cardiac function using TTE. A transesophageal echocardiogram (TEE) could be a better screening tool for this group of patients. The quality of life, based upon a subjective score as mentioned by patients, was a reliable tool to indicate the postoperative relief of symptoms. Medical photography should always be performed before and after surgery as it provides valuable information about the patient's defect.[34] In addition, an objective evaluation of the cosmetic effect can be made. This effect was generally regarded as satisfactory (70%).

Conclusions

Our study has several limitations. In the present study, some patients were suffering from pulmonary or coronary disease. This could have influenced the pattern of symptoms and QoL. To give these results increased validity more investigation on larger groups and measurements in children and young adults should be performed.[36-38]. In this way more and more specialists will become familiar with the positive results of treating SPES and patients will benefit.

SPES exists and is a surgically treatable cardiovascular disease.

REFERENCES

1. Fonkalsrud EW. Open pectus excavatum repairs. Changing trends, lessons learned. One surgeon's experience. World J Surg. 2009;33:180–90.
2. Goretsky MJ, Kelly RE, Croitoru D, Nuss D. Chest wall anomalies. Pectus excavatum and pectus carinatum. Adolesc Med. 2004;15(3):455–71.
3. Cobben JM, Oostra RJ, van Dijk FS. Pectus excavatum and carinatum. Eur J Med Genet. 2014 May 9. pii: S1769-7212 (14)00102-5.
4. Kelly RE, Shamberger RC, Mellins RB, et al. 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. 2007;205:205–216.
5. Creswick HA, Stacey W, Kelly RE, et al. Family study of the inheritance of pectus excavatum. J Pediatr Surg. 2006;41:1699–03.
6. Heithaus JL, Davenport S, Twyman KA, Torti EE, Batanian JR. An intragenic deletion of the gene MNAT1 in a family with pectus deformities. Am J Med Genet A. 2014 May;164(5):1293-7.
7. Dubois-Laforgue D, Bellanne-Chantelot C, Subra JF, Timsit J. Pectus excavatum is part of the clinical spectrum of HNF1B MODY 5. Diabetes Care, 2014 Apr;37(4):e72-3.
8. Jeong JY, Park HJ, Lee J, Park JK, Jo KH. Cardiac morphologic changes after the Nuss operation for correction of pectus excavatum. Pediatr Surg Int (2011) 27:61–66
9. Malek MH, Berger DE, Housh TJ, Marelich WD, Coburn JW ,Beck TW. Cardiovascular Function Following Surgical Repair of Pectus Excavatum A Metaanalysis. Chest 2006;130:506-516
10. Jaroszewski D, Notrica D, McMahon L, Steidley E, Deschamps C. Current Management of Pectus Excavatum. A Review and Update of Therapy and Treatment Recommendations. J Am Board Fam Med 2010;23:230 –239
11. Kim M, Lee KY, Park HJ et al. Development of new cardiac deformity indexes for pectus excavatum on computed tomography. Feasibility for pre- and post-operative evaluation. Yonsei Med J. 2009; 50(3): 385-390
12. Gürkan U, Aydemir B, Aksoy S, Akgöz H, Rodi A et al. Echocardiographic Assessment of Right Ventricular Function Before and After Surgery in Patients with Pectus Excavatum and Right Ventricular Compression. Thorac Cardiovasc Surg. 2014 Apr; 62(3):231-5
13. Croitoru DP, Kelly RE,. Goretsky MJ,Lawson ML, Swoveland B, Nuss D. Experience and modification update for the minimally invasive Nuss technique for pectus excavatum repair in 303 patients. J Pediatr Surg 2002;37: 437-445.

14. Kelly RE. Pectus excavatum. Historical background, clinical picture, preoperative evaluation and criteria for operation. Semin Pediatr Surg. 2008;17:181-93
15. Pectus excavatum not always as harmless as it seems. Winkens R ,Guldemond FI, Höppener PF, Kragten JH, van Leeuwen Y. BMJ Case Reports 2009;doi:10.1136/bcr.10.2009.2329
16. Kragten HA, Siebenga J, Höppener PF, Verburg R, Visker N.Symptomatic pectus excavatum in seniors (SPES): a cardiovascular problem? : A prospective cardiological study of 42 senior patients with a symptomatic pectus excavatum. Neth Heart J. 2011 Feb;19(2):73-8.
17. Jaroszewski D, Steidley E, Galindo A, Francisco Arabia A. Treating heart failure and dyspnoea in a 78-year-old man with surgical correction of pectus excavatum. Ann Thorac Surg. 2009;88:1008–10
18. Crump HW. Pectus excavatum—pathophysiology, clinical presentation, surgical repair. Am Fam Physician. 1992;46:173–9.
19. Jaroszewski DE, Fonkalsrud EW. Repair of pectus chest deformities in 320 adult patients: 21 year experience. Ann Thorac Surg. 2007;84: 429–33.
20. Mansour KA, Thourani VH, Odessey EA, Durham MM, Miller JI. Thirty-year experience with repair of pectus deformities in adults. Ann Thorac Surg. 2003;76: 391–5
21. Krueger T, Chassot PG, Christodoulou M, Cheng C, Ris HB, Magnusson L. Cardiac function assessed by transesophageal echocardiography during pectus excavatum repair. Ann Thorac Surg. 2010;89: 240–4.
22. Nuss D. Recent experiences with minimally invasive pectus excavatum repair (Nuss procedure). Read at the Fifty-seventh Annual Meeting of the Japanese Association for Thoracic Surgery, Educational Lecture, Sapporo, October , 2004.
23. Krasopoulos G, Dusmet M, Ladas G, Goldstraw P. Nuss procedure improves the quality of life in young male adults with pectus excavatum deformity. Eur J Cardiothor Surg 2006; 29:1-5
24. Neviere R, Montaigne D, Benhamed L, Catto M, Edme JL, Matran R, Wurtz A, Cardiopulmonary response following surgical repair of pectus excavatum in adult patients. European Journal of Cardio-thoracis Surgery, Aug 2011, Vol 40, issue 2, pages e77-e82.
25. Humphries CM, Anderson JL, Flores JH, Doty JR., Cardiac magnetic resonance imaging for perioperative evaluation of sternal eversion for pectus excavatum. Eur J Cardiothorac Surg. 2013 Jun;43(6):1110-3.
26. Saleh RS, Finn JP, Fenchel M, Moghadam AN, Krishnam M, Abrazado M, Ton A, Habibi R, Fonkalsrud EW, Cooper CB., Cardiovascular magnetic resonance in patients with pectus excavatum compared with normal controls. J Cardiovasc Magn Reson. 2010 Dec 13; 12:73.
27. Rattan A.S, Laor T., Ryckmanm F.C, Brody A.S, Pectus excavatum imaging: enough but not too much. Pediatr Radiol (2010) 40:168-172.
28. Marcovici P., LoSasso B, Kruk P, Dwek J. MRI for the evaluation of pectus excavatum.Pediatr Radiol(2011) 41: 757-758.
29. Kelly RE Jr1, Mellins RB, Shamberger RC, Mitchell KK, Lawson ML, Oldham KT, Azizkhan RG, Hebra AV, Nuss D, Goretsky MJ, Sharp RJ, Holcomb GW 3rd, Shim WK, Megison SM, Moss RL, Fecteau AH, Colombani PM, Cooper D, Bagley T, Quinn A, Moskowitz AB, Paulson JF. Multicenter study of pectus excavatum, final report: complications, static/exercise pulmonary function, and anatomic outcomes. J Am Coll Surg. 2013 Dec;217(6):1080-9.
30. Maagaard M, Tang M, Ringgaard S, Nielsen HH, Frøkiær J, Haubuf M, Pilegaard HK, Hjortdal VE. Normalized cardiopulmonary exercise function in patients with pectus excavatum three years after operation. Ann Thorac Surg. 2013 Jul;96(1):272-8.
31. Janusz Kowalewski , Stanislaw Barcikowski, Marian Brocki. Cardiorespiratory function before and after operation for pectus excavatum. Mediumterm results. European Journal of Cardio-thoracic Surgery 13 (1998) 275–279
32. Jayaramakrishnan K, Wotton R, Bradley A, Naidu B. Does repair of pectus excavatum improve cardiopulmonary function. Interact Cardiovasc Thorac Surg. 2013 Jun;16(6):865-70.
33. Malek MH , Dale E. Berger DE ,Marelic WD, Coburn JW ,Beck TW, Housh TJ. Pulmonary function following surgical repair of pectus excavatum. A metaanalysis. Eur J Cardiothorac Surg 2006;30: 637-643

34. van Dijk H, Höppener PF, Siebenga J, Kragten HA. Medical photography: a reliable and objective method for documenting the results of reconstructive surgery of pectus excavatum. J Vis Commun Med. 2011 Mar;34(1):14-21.
35. Dawn E. Jaroszewski, MD, MBA, FACS, Tahlil A. Warsame, BSE, RDCS, Krishnaswamy Chandrasekaran, MD, and Hari Chaliki, MD . Right Ventricular Compression Observed in Echocardiography from Pectus Excavatum Deformity. J Cardiovasc Ultrasound. Dec 2011; 19(4): 192–195.
36. Poston PM, Patel SS, Rajput M, Rossi NO, Davis JE, Turek JW. Defining the role of chest radiography in determining candidacy for pectus excavatum repair. Innovations (Phila). 2014 Mar-Apr; 9(2):117-21.
37. Chan Wah Hak YS, Lim YP, Liew R, Hsu LF. Pectus excavatum: uncommon electrical abnormalities caused by extrinsic right ventricular compression. J Cardiovasc Electrophysiol. 2014 Mar; 25(3):324-7.
38. Jaroszewski DE. Physiologic implications of pectus excavatum. J Thorac Cardiovasc Surg. 2017 Jan;153(1):218-219