

TREATMENT OF PECTUS EXCAVATUM



Mustafa YÜKSEL¹
Nezih Onur ERMERAK²

INTRODUCTION

Pectus Excavatum (PE), also known as the funnel chest, is the most common chest wall deformity (1). It is characterized by the depression of the anterior chest wall and sternum. They occur in 1 of every 300 to 400 births and the deformity is seen 5 times more often in males compare to women (2). In a recent publication; Biavati and friends reported that population-based prevalence of pectus excavatum in adults of 0.4%. Leonardo da Vinci portrayed the first image of PE in 1510 which makes it the first ever presentation of the deformity (3). Egge published the first report in the literature in 1870 and identified the deformity as a depression of the anterior chest wall (4). Kwicinski reported the evaluation of computed tomography (CT) scans of 217 mummies in which 3 PE cases were detected. This information reveals the presence of the deformity in antique times with similar prevalence up to date.

Even though the etiology of the deformity is not clearly identified yet, there are some theories and hypothesis remarking the pathophysiology of PE. These theories indicate the posterior traction of the sternum and intrinsic failure of osteochondrogenesis secondary to intrauterine pressure, rickets, pulmonary restriction or diaphragmatic deformities. In addition to these theories, some biochemical irregularities, which include malfor-

mation in the type 2 collagen production in costal cartilages and abnormal levels of zinc, magnesium and calcium, were detected (5).

The genetic perspective of PE still remains unknown, but clinical evidence supports that PE is a genetic disorder as 40% of the patients have affected family members with similar congenital deformities (5). Although we couldn't identify any specific loci in any genes directly related to disorder, there are new studies showing some potential genomes (6,7,8).

Clinically, PE patients have a stereotypical posture: thin, tall patients with a pot-belly and forward-drifted shoulders. Although it is still controversial and there is no consensus among physicians as to whether or not pectus excavatum can produce symptoms enough to justify a surgical procedure, posterior traction of the sternum can displace the heart and may have a negative effect on lung volume and cardiac functions. As a consequence of this anatomical disposition some patients may experience chest pain, dyspnea on exertion, palpitations, fatigue and low exercise tolerance (9,10,11). Dr.Gaston and friends published the data about cardiac morphology and function according to the site of maximum compression and reported that patients with a compression to the right ventricle and AV groove experience diverse cardiac abnormalities (12). In

¹ Prof. Dr., Emeritus Prof. of Marmara University, Lecturer of TC Demiroğlu Bilim University Medical School, drmustafayuksel@gmail.com,

² Asst.Prof. M.D Marmara University;Faculty of Medicine. Department of Thoracic Surgery,Istanbul/Turkey

the surgery can sometimes turn into a long lasting issue and ends up with early unplanned removal of the bar. Satisfaction rates after correction with MIRPE are reported between 80 – 97%. Patients who underwent MIRPE could be able to return their daily life in very short periods of time, in relation to the nature of minimally invasive surgeries and as MIRPE being one of them.

1084 patients had been operated with MIRPE between August 2005 – December 2020 (Resim 1,2.). 937 (86.4%) patients were male, 147 (13.6%) were female and the median age was 18.62 years (range: 4-58). The deformity was symmetric in 824 (76%) and asymmetric in 260 (24%) cases. One pectus bar was used in 661 (61%) cases, two in 401 (37%), three in 22 (2%) cases for the correction of the deformity. The median operation duration was 72 minutes (range: 20-180) and the median duration of hospital stay was 4 ± 1.5 days (range: 2-10). Scoliosis was the most common co-existing anomaly with 252 (23.2%).

176 (16.2%) patients had a positive familial history for chest wall deformities. Postoperative management was maintained by epidural analgesia in 85 (7.8%) patients and PCA in 999 (92.2%) patients. Quality-of-life (QOL) questionnaires revealed 95% satisfaction in terms of correction.

In concordance with the existing literature; as seen in 38 (3.5%) patients pneumothorax emerges as the most common perioperative complication. While 8 of them needed chest tube insertion, the rest of them were treated conservatively. Pleural effusion was detected in 9 (0.83%) patients whom only 3 drained with tube thoracostomy . Thoracotomy was performed in 2 (0.18%) patients owing to myocardial laceration.

Wound infections and insufficient correction were the leading cause of late postoperative complications with 56 (5.1%) and 20 (1.8%), respectively. 9 of the patients with insufficient correction were managed with second surgery for the correction. Cellulitis (8 patients 0.73%), bar displacement (8 patients 0.73%) , seroma (3 patients -0.35%) and pneumothorax (5 patients 0.46%) were the other late postoperative complications.

Bar removal was performed in 764 patients, 622 of whom were on scheduled time frame. Most alarming complication during bar removal surgeries was life threatening hemorrhage which managed by packing conservatively in 3 of them. While one of the other two needed left thoracostomy, extension of the primary incision was enough in the other one.

REFERENCES

1. Brochhausen C, Turial S, Muller FK, Schmitt VH, Coerdet W, Wihlm JM et al. Pectus excavatum: history, hypotheses and treatment options. *Interact CardioVasc Thorac Surg* 2012;14:801–6.
2. Jaroszewski D, Notrica D, McMahon L, Steidley DE, Deschamps C. *Current Management of Pectus Excavatum: A Review and Update of Therapy and Treatment Recommendations*. *J Am Board Fam Med* 2010;23(2):230- 9
3. Ashrafian H. Leonardo da and the first portrayal of pectus excavatum. *Thorax* 2013;68:1081.
4. Eggel. *Eine seltene Mißbildung des Thorax*. *Virchows Arch Path Anat*. 1870;49:230
5. Creswick HA, Stacey MW, Kelly Jr RE, Gustin T, Nuss D, Harvey H, Goretsky MJ, Vasser E, Welch JC, Mitchell K, Proud VK. *Family Study of the Inheritance of Pectus Excavatum*. *J Pediatr Surg* 2010 ;41(10):1699-1703
6. Wu S, Sun X, Zhu W, Huang Y, Mou L, Liu M, Li X, Li F, Li X, Zhang Y, Wang Z, Li W, Li Z, Tang A, Gui Y, Wang R, Li W, Cai Z, Wang D. Evidence for GAL3ST4 mutation as the potential cause of pectus excavatum. *Cell Research* 2012;22:1712-1715
7. Karner CM, Long F, Solnica-Krezel L, Monk KR, Gray RS. Gpr126/Adgrg deletion in cartilage models idiopathic scoliosis and pectus excavatum in mice. *Hum. Mol. Genet* 2015;24(10):4365-4373
8. Tong X, Li G, Feng Y. TINAG mutation as a genetic cause of pectus excavatum. *Med. Hypotheses* 2020;137:109557
9. Fonkalsrud EW. Current management of Pectus Excavatum. *World J. Surg* 2003;27:502508
10. Goretsky MJ, Kelly RE, Croitoru D, Nuss D. Chest wall anomalies: pectus excavatum and pectus carinatum. *Adolesc Med* 2004;15:455-471
11. Koumbourlis AC. Pectus Excavatum: Pathophysiology and clinical characteristics. 2009;10:3-6
12. Rodriguez-Granillo GA, Raggio IM, Deviggiano A, Munzon-Bellia G, Capunay C, Nazar M, Martinez JL, Carrascosa P, Martinez-Ferro M. Impact of pectus excavatum on cardiac morphology and function according to the site of maximum compression: effect of physical exertion and respiratory cycle. *Eur Heart J Cardiovasc Imaging* 2020;21(1):77-84
13. Cheng YL, Lan CC, Wu YK, Su WL, Yang MC. Poorer sleep quality among adult patients with pectus excavatum in Taiwan: A pilot study. *J Thorac Cardiovasc Surg* 2019;157:769-80

14. Rachwan RJ, Purpura AK, Kahwash BM. Sudden Cardiac Arrest in a Young Patient With Severe Pectus Excavatum. *Am. J.M.Sc* 2018;356(6):570-573
15. Jaroszewski D, Steidley E, Galindo A, Arabia F. Treating Heart Failure and Dyspnea in a 78-Year-Old Man with surgical correction of Pectus Excavatum. *Ann. Thorac. Surg.* 2009;88(3):1008-1010
16. Ravitch MM. The operative treatment of pectus excavatum. *Ann Surg.* 1949;129(4):429-444.
17. Nuss D, Kelly RE Jr, Croitoru DP, et al. A 10 year review of a minimally invasive technique for the correction of pectus excavatum. *J Pediatr Surg.* 1998;33:545-552.
18. Chavoïn JP, Grolleau JL, Moreno B, Brunello J, Andre A, Dahan M, Garrito I, Chaput B. Correction of Pectus Excavatum by Custom-Made Silicone Implants: Contribution of Computer-Aided Design Reconstruction. A 20-Year Experience and 401 cases. *Plast. Reconstr. Surg.* 2016;137:860-871
19. Haecker FM. The vacuum bell for conservative treatment of pectus excavatum: the Basle experience. *Pediatr Surg Int* 2011;27:623-7.
20. Obermeyer RJ, Cohen NS, Kelly RE, Kuhn MA, Frantz FW, McGuire MM, Paulson JF. Non-operative management of pectus excavatum with vacuum bell therapy: a single-center study. *J Pediatr Surg* 2018;53:1221-1225
21. Jayaramakrishnan K, Wotton R, Bradley A, Naidu B. Does repair of pectus excavatum improve cardiopulmonary function? *Interct Cardio Th* 2013;16:865-871
22. Nevieré R, Montaigne D, Benhamed L, Catto M, Edme JL, Matran R, Wurtz A. Cardiopulmonary response following surgical repair of pectus excavatum in adult patients. *Eur J Cardiothorac Surg* 2011;40:77-82
23. Maagaard N, Tang M, Ringgaard S, Nielsen HHM, Frokiaer J, Haubuf M, Pliegaard HK, Hjortdal VE. Normalized Cardioplumonary Exercise Function on Parient with Pectus Excavatum Three Years After Operation. *Ann Thorac Surg* 2013;96:272-278
24. 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(2):506-516
25. Malek MH, Berger DE, Marelich WD, Coburn JW, Beck TW, Housh TJ. Pulmonary function following surgical repair of pectus excavatum: A meta-analysis. *Eur J Cardiothorac Surg* 2006;30: 637-643
26. Guntheroth WG, Spiers PS. Cardiac function before and after surgery for pectus excavatum. *Am J Cardiol* 2007;99:1762-1764
27. Johnson JN, Hartman TK, Pianosi PT, Driscoll DJ. Cardiorespiratory function after operation for pectus excavatum. 2008;153:359-364
28. Aronson DC, Bosgraaf RP, Merz EM, Van Steenwijk RP, van Aalderen WMC, van Baren R. Lung function after the minimal invasive pectus excavatum repair (Nuss procedure). *World J Surg* 2007 ;3 (7);1518-1522
29. St.Peter SD, Juang DJ, Garey CL, Laituri CA, Ostlie DJ, Sharp RJ, Snyder CL. A novel measure for pectus excavatum: the correction index. *J Pediatr Surg* 2011;46:2270-2273
30. Sesia SB, Heitzelmann M, Schaedelin S, Magerkurth O, Kocher GJ, Schmid RA, Haecker FM. Standardized Haller and Asymmetry index combined for a more accurate assessment of pectus excavatum. *Ann Thorac Surg* 2019;107:271-6
31. Daunt SW, Cohen JH, Miller SF. Age-related normal ranges for the Haller Index in children. *Pediatr Radiol* 2004;34(4):326-30
32. Kelly RE Jr. *Pectus excavatum: historical background, clinical picture, preoperative evaluation and criteria for operation. Semin Pediatr Surg.* 2008;17(3):181-93.