



Minimally invasive repair of pectus excavatum in a 17-year-old boy with a history of congenital diaphragmatic hernia and lack of pericardium



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ABSTRACT

We reported a 17-year-old boy with very deep asymmetric pectus excavatum and with a history of congenital diaphragmatic hernia repair and hypoplastic left lung. We performed a minimally invasive repair of pectus excavatum as described by Nuss et al., in 1998. We performed a left-sided thoracoscopy, instead of the right-sided according our normal routine, to provide a safe route. We created a substernal tunnel to have a clear definition of the deviant anatomy after congenital diaphragmatic hernia repair. However, we noticed an absence of the pericardium, which, by itself, can increase the risk of cardiac injury in both bar insertion and removal. Instead of the usual right-sided thoracoscopy, we recommend providing a safe view by left-sided thoracoscopy in comparable cases (e.g. congenital diaphragmatic hernia, other cardiac or vascular malformations) to reduce the risk of rupture or perforation of cardiac structures.

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Pectus excavatum (PE) or ‘funnel chest’ is by far the most common congenital thoracic wall deformity which affects 1/400 children and is predominantly found in male patients [1,2]. PE can be treated either by the ‘modified Ravitch procedure,’ an open surgical procedure with resection of the deformed costal cartilage and sternal osteotomy, or by ‘minimally invasive repair of pectus excavatum’ (MIRPE) also, called the ‘Nuss-procedure,’ by placing a retrosternal bar to uplift the sternum. The first insertion of a retrosternal stainless steel bar was published in 1975 by Skobos et al. This steel bar was added to the conventional open procedure to prevent relapse of the sternum [3]. In 1998, Nuss et al., published their first results of correcting PE by placing a retrosternal stainless steel bar without performing costal cartilage resection or sternal osteotomy as an answer to the labor-intensive conventional Ravitch procedure [4]. MIRPE has excellent cosmetic results and a low complication rate [5–7]. Only a few major complications, such as cardiac perforation, hemothorax, pulmonary vessel erosion, are reported in literature [8–12]. To minimize the risk of mediastinal or cardiac injury, most of the surgeons perform MIRPE thoracoscopy-guided [13]. There have been two reports of minimally invasive

repair for pectus excavatum done in a patient with a history of congenital diaphragmatic hernia in literature in the past [14]. We report our altered technique for MIRPE in a 17-year-old boy with a very deep pectus excavatum and a history of congenital diaphragmatic hernia repair, hypoplastic left lung by left-sided thoracoscopy instead of the more commonly used right-sided thoracoscopy.

1. Case report

A 17-year-old boy with a history of congenital diaphragmatic hernia and hypoplastic left lung visited our clinic three years ago. The patient underwent diaphragmatic hernia repair one-week post partum. In the following years, the patient developed a pectus excavatum with asymmetric anterior wall in disadvantage of the left thoracic wall and a scoliosis. The patient had no physical complaints. A cardiac magnetic resonance imaging (CMR) was performed and showed a very deep asymmetric pectus with a Haller index (HI) of 16 and a mispositioned heart in the left thorax (Fig. 1).

The minimal distance between sternum and the spinal cord was less than 1 inch. Compression of the right atrium and ventricle and elongation of the great vessels was described. However, no evidence for flow obstruction, malfunction of the valves was witnessed. The left and right ventricle function seemed to be normal.

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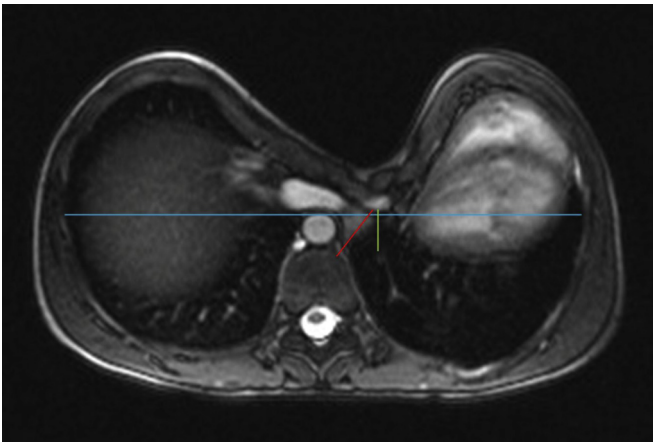


Fig. 1. CMR shows a very deep asymmetric PE and a mispositioned heart in the left thorax. Red line: minimal distance between spine and sternum <1 inch. Haller index: 16.

2. Bar insertion technique

We performed a minimally invasive repair of pectus excavatum, video-assisting thoracoscopic surgery (VATS) guided and monitored by electrocardiography (ECG). We implanted a 12-inch bar. Due to the abnormal anatomy, the thoracoscope (30°) was first inserted into the left thorax to avoid cardiac, vascular or pulmonary injury. The trocar was placed in the fourth intercostal space in the anterior axillary line. In our opinion is left-sided introduction of the first trocar safe. Firstly, the fourth intercostal space in the anterior axillary line is above the heart. Secondly, after carefully opening of the pleural space the lung will collapse and the heart will shift backwards to the posterior thoracic wall. Unexpectedly, we noticed the absence of the pericardium during inspection of the left thorax (Fig. 2).

Furthermore, we safely introduce to pectus introducer in the left thoracic space. A left-to-right substernal tunnel was created away from the heart instead of toward the heart. Due to the extreme deep pectus we were unable to bring the pectus introducer to the right-sided extra-thoracic tunnel. Therefore, we created a small extra-

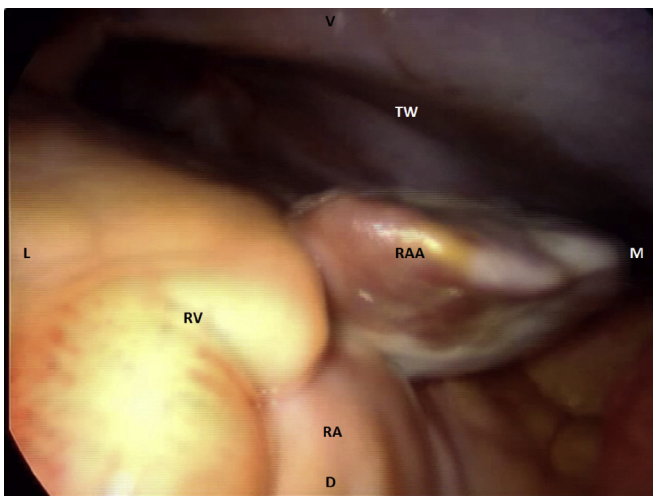


Fig. 2. Thoracoscopic left-sided view shows cardiac structures in absence of pericardium. Anatomic position: L = lateral, M = medial, V = ventral, D = dorsal, structures, RV = right ventricle, RA = right atrium, RAA = right atrial auricle/appendage, TW = thoracic wall.

right-sided incision and used a steel wire to lift the pectus introducer to advance it through the thoracic cavity. The use of substernal lifting was unattractive due to his former CDH repair which crosses the linea alba. Bar insertion went without further complications. There were no events during follow-up. Fig. 3 shows pre- and post-operative results.

3. Bar removal technique

Three years later the bar was removed during an elective procedure. We expected a higher risk of cardiac adhesions to the thoracic wall or to the Nuss bar because of the lack of its natural cardiac sheet (i.e. pericardium). Consequently, we had VATS standby in case any arrhythmias were shown on ECG which may occur during bar manipulation due to cardiac adhesions. The bar removal went without complications. After surgery, the patient fully recovered and was discharged the same day.

4. Discussion

There is minimal experience of MIRPE in patients with CDH. Theoretically, these patients have an increased risk of major complications during bar placement such as cardiac or pulmonary rupture or perforation due to formed adhesions in the thoracic cavity after CHD repair in childhood. Moreover, extremely severe pectus excavatum, such as in our case, seems to have a higher risk on major complications such as perforation of cardiovascular structures [10,15,16]. Kabbaj et al., recommend a sub- and retro-xiphoid approach to guide implant insertion or a classic sternochondroplasty procedure instead of thoracoscopic PE correction in this situation [17]. In addition, lack of pericardium layer to protect the heart and great vessels aggravates the risk of perforation during bar insertion as well as during bar removal [18]. The combination of CDH repair, severe PE and absence of pericardium makes this a high risk case for MIRPE. We advise providing a safe view by a left-sided thoracoscopy. The left-sided view will ensure a safer way to create a substernal tunnel and to insert the bar without damaging any cardiovascular structures. In addition, Metzelder et al., also describe a safe thoracoscopic adhesiolysis before bar placement by left-sided thoracoscopy [14]. Furthermore, a lack of pericardium was unexpected in our case. Apparently, it is a very rare anomaly with an incidence of less than 1/10,000 [19,20]. There are only a few cases of congenital absence of the pericardium associated with PE reported in the literature [21]. Its association with CDH remains unclear. CMR is considered the gold standard to detecting absence of pericardium [22]. Unfortunately, even after revision we could not recognize it on CMR in our case. For bar removal, lack of pericardium increases the risk of cardiovascular adhesions to the thorax wall or retrosternal bar. Therefore, we advise removing the bar guided by ECG or VATS. ECG, in this case, provides more information about arrhythmias during manipulation of the bar caused by adhesions between cardiac structures and the bar. Our ECG did not show any arrhythmias during careful manipulation. Therefore, new cardiac adhesions were unlikely, so we did not require use of VATS during removal. For bar removal, we intended to use VATS for an optimal view just as in the previous procedure (cause of absence of cardiac protection layer: pericardium). In conclusion, we provided a very safe and clear view for MIRPE by left-sided thoracoscopy in a case of severe PE, CHD repair. Furthermore, we did a successful ECG-guided bar removal, despite the lack of pericardium (i.e. with an increased risk of cardiovascular adhesions to the thorax wall or retrosternal bar). In order to reduce the risk of rupture or perforation of cardiac structures we recommend ECG and, if the ECG shows any arrhythmias during traction, left-sided VATS for a safer removal in case of any cardiac adhesions.



Fig. 3. Left image: pre-operative status. Note the scar after CDH repair. Right image: post-operative result after placement of a single bar.

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