

Evolution in the management of pectus excavatum in pediatric patients

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Comment on: Scalise PN, Demehri FR. The management of pectus excavatum in pediatric patients: a narrative review. Transl Pediatr 2023;12:208-20.

Keywords: Pectus excavatum; pediatric patients; Nuss procedure; vacuum bell; cryoablation

Submitted Apr 26, 2023. Accepted for publication Jul 19, 2023. Published online Jul 28, 2023. doi: 10.21037/tp-23-264 View this article at: https://dx.doi.org/10.21037/tp-23-264

The majority of congenital chest wall deformities (CWD) affects the anterior chest wall. The most common anterior CWD is pectus excavatum (PE), followed by Pectus carinatum (PC). The incidence of PE is approximately 1 in every 300–400 births, affecting male patients with an approx. 4:1 ratio (1). For the last 70 to 80 years, open surgical repair such as the Ravitch technique and its modifications (2,3) was the preferred method to correct CWDs. An essential paradigm shift occurred with the inauguration of the minimally invasive repair of pectus excavatum (MIRPE) by Nuss (4). In contrast to open repair, MIRPE does not require cartilage or sternal resection. Flexibility and elasticity of the chest wall which are preserved applying MIRPE, were identified as relevant parameters for successful treatment of CWD. Furthermore, an increasing number of patients presented with thoracic chondrodystrophy as a long-term side effect after open surgical repair. As a consequence, the management of CWD including diagnostic work-up as well as conservative and surgical treatment options has made substantial progress during the past two decades. Today, treatment of CWD includes all age groups with pediatric, adolescent and adult patients, covered by a dedicated multidisciplinary team (in particular pediatric and thoracic surgeons) based in a specialized chest wall unit, what is the most important pillar

of present time.

The physiologic effects of CWD are still discussed controversially. There is an ongoing controversial debate concerning effects of MIRPE on exercise tolerance, lack of endurance, shortness of breath, cardiopulmonary function, body posture, etc. Even if the number of articles reporting on different aspects of CWD treatment went up from approximately 300 (1980 to 1989) to more than 1,000 published papers (2012 to 2021), the controversial debate will continue (5). Information on new diagnostic and therapeutic modalities provided by online platforms and social media circulates not only among surgeons and paediatricians, but also rapidly among patients. Not only different surgical techniques, but also conservative treatment options are available. In almost the same manner, postoperative pain management is an important part of an effective and successful treatment protocol. Several modifications were introduced over the past years. For a long time, regional analgesia such as paravertebral nerve blocks or catheters, epidurals as well as patient-controlled analgesia (PCA) represented the preferred anesthesia method. Nowadays, cryoablation became a more and more applied alternative technique (6,7). Cryoablation may be applied thoracoscopically during MIRPE (8), or as ultrasound guided percutaneous intercostal injection (9).

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Translational Pediatrics, Vol 12, No 8 August 2023

After its introduction, MIRPE was well established in the first decade of this century and subsequently performed with increasing frequency worldwide. Today, MIRPE represents the worldwide used "gold-standard" for surgical repair of PE. Applying the technique in different age groups, the method experienced numerous modifications. Modifications include patient selection and indication, preoperative evaluation, as well as age at time of surgical repair. Positioning of the patient on the OR table, intubation (single lumen vs. double lumen tubus) were modified. Number, location and size of skin incisions, shape/size or number of bars and bar fixation as well as bar passage (intra vs. extrapleural placement) were modified (10). To reduce the risk of secondary bar displacement and/or to correct complex CWDs, placement of more than 1 bar including the so called cross-bar technique was introduced (11).

Two fundamental techniques were introduced to reduce the risk of intraoperative cardiac injuries: routine unilateral and/or bilateral thoracoscopy (12), and routine sternal elevation (13,14). Whereas in the initial publication by Nuss retrosternal dissection was described as "blunt" using a long-curved clamp without thoracoscopy (4), the same group reported their experience using routine thoracoscopy 4 years later, achieving improved safety during mediastinal dissection (15). In particular the risk of cardiac perforation as the most severe complication could be decreased with the routine use of thoracoscopic guidance (16). Even more than 2 decades after its introduction there is no evidence-based data available concerning the protective effect of thoracoscopy on the true incidence of near fatal complications like cardiac injuries. However, the majority of articles and studies reporting on catastrophic complications were all published before 2011 (5). Furthermore, the widespread use of routine intraoperative sternal elevation must be considered as an additional effective measure to improve safety of MIRPE (13). Visualization across the mediastinum during retrosternal dissection may be compromised in severe defects. Notably in older adult PE patients, decreased flexibility and stiffness of the anterior chest wall as well as the corresponding force required to elevate the sternum may make retrosternal dissection difficult and sometimes nearby impossible. Applying routine sternal elevation during MIRPE, the safety of the procedure has improved clearly as there was no near-fatal and/or fatal incident reported anymore when the technique was applied intraoperatively (13).

Evolution in the management of congenital CWD and

modifications of newly introduced techniques have made significant progress over the past 2-3 decades, and as a consequence patients outcome improved substantially. The aim of the narrative review presented by Scalise and Demehri (17) is to outline current practice concerning diagnostic work-up, conservative and surgical treatment as well as management in general of pediatric PE patients. Since we may find numerous publications dealing with this topic, it is nearby impossible to review all the literature, covering all different age groups. In particular no randomized double-blind studies are available. Therefore, the quality of evidence regarding this topic remains relatively low with still many unexplored and unknown pathways. Different objective criteria were screened, verified and added to identify PE patients that would benefit from surgery. An increasing number of studies accentuating the cardiopulmonary consequences of PE. Notably, more than 275 papers have been published in the last 10 years focusing on functional changes prior to and after surgical correction of PE. Due to different reasons (e.g., retrospective and/or small cohort studies), the results are still heterogenous and of low evidential quality (18). Based on this generally poor evidence, the controversial debate on the cardiopulmonary impact of PE and whether patients may benefit from surgical repair or not will continue as long as we may be able to provide double blind randomized studies. However, IRB approval for such a study has to be considered as difficult or nearby impossible. This is aggravated by the fact that PE is many times considered as an "only" esthetic disorder and in particular associated with body image disturbances. In contrast, among surgeons and confirmed by recently published studies it is clearly acknowledged that subjective improvement in exercise intolerance is often reported after MIRPE (19). Thus, despite above mentioned facts as well as current believes, reviews such as presented by Scalise and Demehri may help to bridge the lack of convincing high-quality evidence which remains a critical concern that could potentially interfere with future management of PE. As concluded by Scalise and Demehri in their narrative review, there is still a lack of international consensus guidelines concerning preoperative diagnostic work-up, conservative and surgical treatment as well as postoperative management. A recently published study by Janssen et al. reported on a consensus statement for perioperative care for PE based on a multi-round Delphi survey (20). Pectus surgeons all around the world are encouraged to draw up standardized consensus guidelines to establish a high-level algorithm for preoperative diagnostic

Haecker. Management of pectus excavatum in pediatric patients

protocol, indications for conservative and/or surgical treatment as well as postoperative management.

Acknowledgments

Funding: None.

Footnote

Provenance and Peer Review: This article was commissioned by the editorial office, *Translational Pediatrics*. The article did not undergo external peer review.

Conflicts of Interest: The author has completed the ICMJE uniform disclosure form (available at https://tp.amegroups.com/article/view/10.21037/tp-23-264/coif). The author has no conflicts of interest to declare.

Ethical Statement: The author is accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

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1452

Translational Pediatrics, Vol 12, No 8 August 2023

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Cite this article as: Haecker FM. Evolution in the management of pectus excavatum in pediatric patients. Transl Pediatr 2023;12(8):1450-1453. doi: 10.21037/tp-23-264

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