Advancing pectus deformity care: evaluation of current treatments, complications and future innovations

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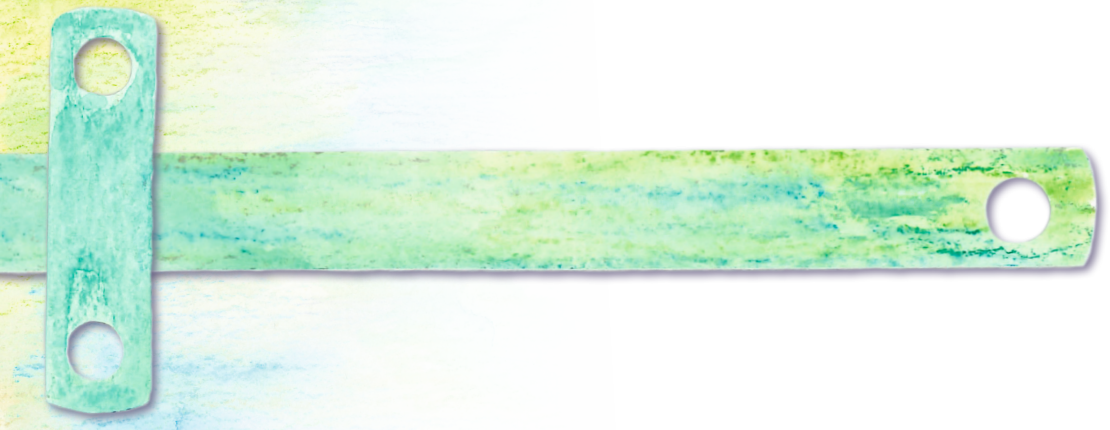

Chapter 1

General introduction and thesis outline

GENERAL INTRODUCTION

This thesis is about pectus deformities. Such chest wall conditions were recognized as early as antiquity, with reports on pectus excavatum dating back to the sixteenth century.[1] Pectus deformities are typically classified into three types: pectus carinatum (PC), or pigeon breast; pectus excavatum (PE), also known as funnel chest; and pectus arcuatum (PA). Each type requires distinct treatment methods.

Pectus carinatum

PC is characterized by the overgrowth of costal cartilage resulting in anterior protrusion of the sternum. It has an incidence ranging from 0.04% to 0.7% in the current literature, with a higher prevalence in males.[2] Although patients are typically asymptomatic, PC is strongly associated with body image disturbance and reduced quality of life.[3-5] Therefore treatment is primarily initiated to enhance body image and quality of life, and for cosmetic reasons.

Treatment

Initially, PC was treated using a traditional, invasive surgical approach; the Ravitch procedure which was first introduced in 1960 and has been modified over the years.[6, 7] Later on, the minimally invasive Abramson procedure was introduced, which is a 'reverse' Nuss procedure. After introduction, this treatment method gradually became the surgical treatment of first choice.[8]

Despite the effectiveness of above surgical options, physicians began to explore non-invasive alternatives early on. Already from the seventies onwards, orthotic treatment using a brace was introduced as an alternative for surgery, with success rates ranging from 40-90%. In addition to the varying success rate, bracing was often complicated by long treatment duration and complications such as discomfort, pain, and skin related problems.[9-13]

In 2008, the Dynamic Compression System brace (DCS-bracing) was introduced.[14] Unlike previous braces, initial research showed DCS-bracing to be highly effective, with reported success rates of up to 90%, with minimal complications, leading to its rapid adoption.[15]

Current challenges

Despite these promising initial outcomes, researchers have not yet compared the results of invasive surgical correction with non-invasive DCS-bracing. Moreover, data on the impact of DCS-bracing on quality of life is limited, with only a few studies addressing this aspect. [12, 16, 17] However, these studies used different types of braces, making comparisons challenging. The limited research on the impact of bracing on quality of life is surprising, given that this is the primary reason patients seek treatment.

Furthermore, it is difficult to determine objective outcomes after bracing since PC is mainly a cosmetic deformity, in contrast to PE. Therefore outcomes ultimately hinge on the patients' own view of success and satisfaction.[18, 19] Clinicians and patients are likely to have differing opinions on what constitutes a successful correction of PC. Differences in such treatment success expectations between patients and physicians can influence treatment decisions. Yet, to date those differences have not been investigated.

Using objective measurements like 3D scanning for monitoring PC offers significant advantages in both clinical decision-making and patient compliance. By providing clear, measurable data on chest wall deformity through the (external) Haller Index ((E)HI),[9, 20, 21] 3D scanning enables clinicians to track treatment progress without radiation exposure. Additionally, having objective data helps patients see their own progress, which can motivate adherence to non-surgical treatments like bracing. However, for these measurements to be fully effective, there is a critical need for reference values that define normal and abnormal (E)HI ranges across age groups and genders. Reference values allow clinicians to set realistic treatment goals and provide benchmarks for success, making it easier to evaluate the impact of interventions and improving consistency in care. These reference values can also be valuable for assessing treatment outcomes in vacuum bell (VB) therapy for pectus excavatum (PE).

Pectus excavatum

PE is characterized by a sunken sternum due to rib cartilage overgrowth. It has an estimated incidence of 0.1-0.8%.[22] While most studies indicate that PE is more common in males, recent literature suggests it may actually be more prevalent in females.[23] However, many females do not seek medical attention, leading to substantial underreporting in this group. Many adult patients experience physical symptoms such as exercise intolerance, chest pain, and respiratory difficulties, though these symptoms often do not appear until puberty.[23, 24] Patients often realize only after treatment that their exercise tolerance had actually been lower beforehand and improved after treatment, though they were not fully aware of the limitation initially. Most pediatric patients with PE are asymptomatic. Primary reasons for pediatric patients to seek

treatment are psychological distress, body image concerns, and reduced quality of life.[4, 25]

Treatment

Currently, minimally invasive repair of PE (MIRPE), also known as the Nuss procedure, is the standard surgical treatment for PE. MIRPE offers excellent long-term outcomes. Numerous techniques and materials are available for the Nuss procedure, leading to considerable variation in practice.[26] In our clinic in Amsterdam, we are using the short bar technique as described by Pilegaard.[27]

An alternative, non-surgical treatment option is VB therapy, which is usually used in patients with relatively mild deformities.[28]

Current challenges

While the surgical technique for the Nuss procedure is well established, postoperative pain management remains a challenge. Pain management following the Nuss procedure is traditionally handled with continuous epidural analgesia (CEA), but intercostal nerve cryoablation has emerged as a promising alternative, potentially reducing opioid use and hospital stay duration.[29] Despite promising results of intercostal nerve cryoablation, long-term data on complications like neuropathic pain remain sparse and other complications potentially related to cryotherapy, such as pleural effusion and hyperinflammatory reactions following cryotherapy, remain underdiscussed in the literature.[30-32]

In addition to the challenges with pain management, complications such as dislocations and infections remain a concern. Dislocations can be reduced by adapting the surgical technique. In case of an postoperative infection, the Nuss bar remains in situ in most instances, but affected patients often require extended antibiotic therapy and, occasionally, additional surgical interventions. Furthermore, material allergies related to the Nuss bar can sometimes be misdiagnosed as infections, leading to complicated clinical courses. The current literature shows no clear consensus on the choice of antibiotics or the optimal duration of treatment.[33-36]

Pectus arcuatum

Pectus arcuatum (PA) is a distinct condition that combines features of both PC and PE, but differs fundamentally in origin. Unlike PE and PC, which are caused by rib cartilage overgrowth, PA results from the premature fusion of the sternal sutures. This leads to an anterior protrusion of the upper sternum, while the lower sternum is typically neutral or indented. Additionally, the sternum in PA is abnormally short.[37]

Treatment

A Ravitch procedure is necessary to correct this complex deformity, PA cannot be corrected with non-invasive treatment.[37]

Cross-cutting perspectives

An additional challenge in the management of pectus deformities lies in determining the optimal timing of treatment. While early intervention is often recommended to take advantage of chest wall flexibility in adolescence, questions remain about how age and psychological development influence patients' motivation to undergo surgery. During periods of prolonged surgical waiting times, particularly during the COVID-19 pandemic, it was observed that a notable number of patients who reached adulthood while awaiting treatment ultimately declined surgery. These observations raise questions about the potential influence of waiting time, increasing cognitive maturity, evolving self-image, and shifting personal priorities on patients' treatment decisions. Despite its clinical relevance, this topic has not yet been studied.

Aims

The aims of this thesis are to present the long-term outcomes of treatment for both PE and PC, establish a foundation for further innovations in bracing therapy, provide guidance for managing postoperative complications following the Nuss procedure and explore factors influencing treatment decision-making in the context of prolonged waiting times.

THESIS OUTLINE

Part I explores treatment options and long-terms results for PC. **Chapter 2** compares results of DCS-bracing to Ravitch surgery, advocating DCS-bracing as the preferred first-line treatment. **Chapter 3** examines the impact of DCS-bracing on quality of life in patients with PC, demonstrating a positive effect on patients' quality of life. **Chapter 4** addresses the challenge of assessing treatment outcomes for DCS-bracing by evaluating intra- and interobserver agreement on treatment outcomes and pre-treatment severity, highlighting the need for objective outcomes measures. **Chapter 5** builds on these recommendations, investigating the use of 3D scanning to establish reference values for a normal chest wall by comparing 3D scans of healthy children with those of patients with PC. These reference values offer an objective benchmark for assessing treatment success.

Part II examines treatment options for PE and addresses pain management and the management of complications associated with the Nuss procedure. **Chapter 6** outlines the long-term outcomes of VB therapy, highlighting that longer treatment duration and extended daily use are key factors for success. It demonstrates that older patients with a stiffer chest wall can also achieve positive results if highly motivated and compliant. The chapter also shows that VB therapy remains challenging in women. **Chapter 7** compares intercostal nerve cryoablation with continuous epidural analgesia, demonstrating it as a superior analgesic method. **Chapter 8** focuses on hyperinflammatory pleural response resembling complex pneumonia following the Nuss procedure, proposing a guideline for diagnosis and treatment. **Chapter 9** provides an overview of postoperative infections and allergies following the Nuss procedure and presents a novel guideline for their diagnosis and management.

Part III presents **Chapter 10**, which reports an investigation of the impact of waiting times on pediatric patients' decisions to decline pectus surgery. It shows that over half of patients withdrew after prolonged waiting, with younger age and absence of physical symptoms at waitlist entry as predictors. Main reasons included increased body acceptance and physical development through strength training or weight gain, underscoring the need for reconsideration of current treatment strategies, including the integration of psychological counseling.

Part IV provides a summary of the thesis and discusses future directions and advancements in the field of pectus deformities.

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