The Nuss Procedure

Clinical Benefits to Minimally Invasive Repair of Pectus Excavatum

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Pectus Excavatum

Healthy growth and development in children is strengthened by an active life. When patterns of a healthy lifestyle are established in young ages, the benefits are carried forward for the rest of their lives. Some of those benefits translate in tangible outcomes such as stronger bones, muscles and joints, better posture and balance, healthier weight range, etc. Some benefits are connected to better social and psychological development, providing young adults with higher self-esteem, more effective social skills and better performance in school. Pectus excavatum (PE) is a congenital deformity characterized by a symmetrical or asymmetrical inward depression or indentation of the anterior thoracic wall that can be present with varied degrees of torsion of the sternum (Figure 1).[1-3]

Pectus excavatum occurs in approximately 1 in 400 births, by a 4:1 male to female ratio.[4,5] The exact causes of PE are not fully understood, however, genetics seem to be an important factor to consider, as well as correlation with other etiologies including scoliosis, kyphosis, and connective tissue disorders such as Marfan syndrome.[6] Historical perception has labeled PE as a primarily "mild cosmetic disorder" without real physiological effects and should therefore be left alone without surgical intervention.[7] However, "mild" may not be the correct word to describe complaints of chest and back pain, limitations to endure and excel in sports, embarrassment, isolation, anxiety, depression and social awkwardness some of these patients experience. In a matter of weeks or months, what often appeared to be a minor divot around the sternum can become a profound indentation that may affect every aspect of life, that turn around the time of puberty into a life-altering condition with cardiovascular, pulmonary, and psychosocial impairment.[3]

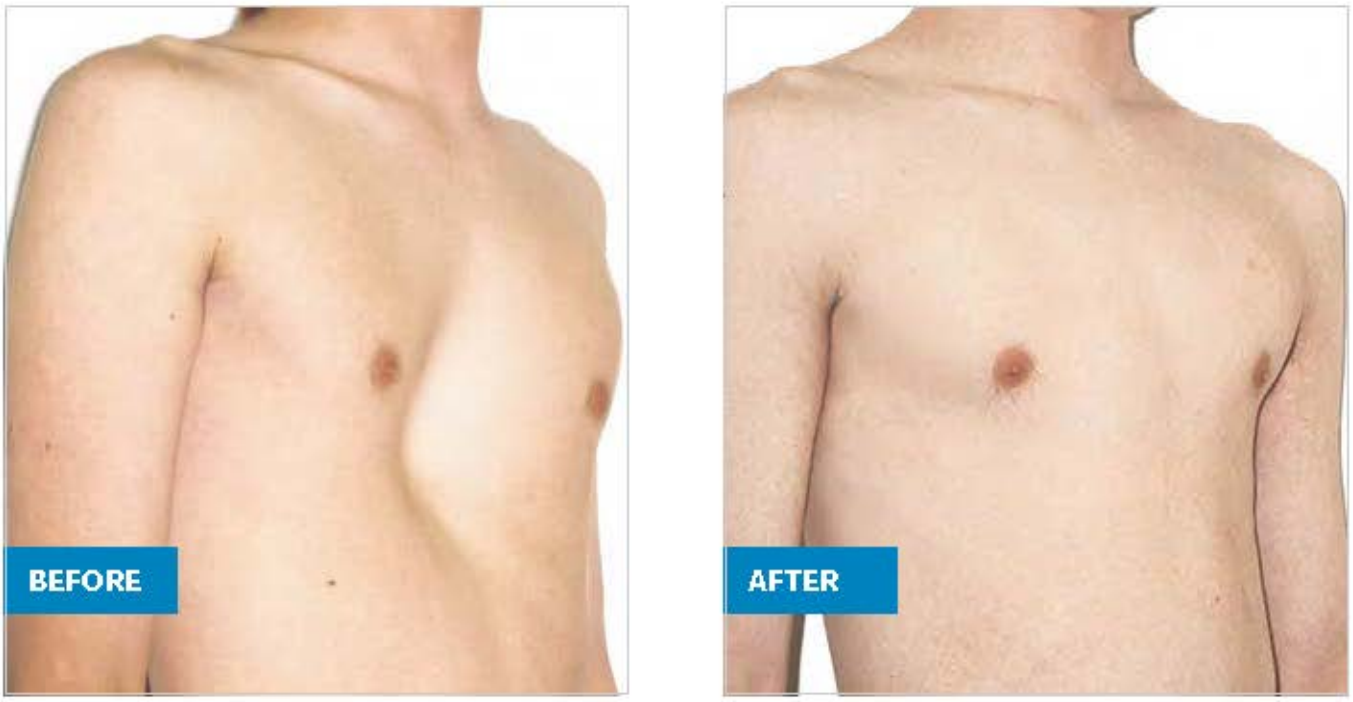

Figure 1: Adolescent presenting with pectus excavatum deformity (Before) showing indentation of the anterior chest wall and same patient showing normal chest wall shape after procedure to correct the deformity (After).

More Than a Cosmetic Defect

Pectus excavatum occurs on a continuum from mild in early stages of life to severe in young adulthood in some cases. The course of pectus excavatum can include symptoms that affect all aspects of life including physical and mental well-being (Figure 2). It manifests with physical symptoms that include varying levels of chest and back pain, pulmonary restriction (both at rest and on exertion), decreased stamina and endurance during exercise, and noticeable posture alterations involving kyphosis of the thoracic spine, forward-sloping shoulders, and a protuberant abdomen.[8-10] Cardiac displacement and restrictive cardiomyopathy occur with more severe deformities, although these and other pulmonary signs due to volume restriction show up readily on CT scans.[9] Patients with pectus may also experience frequent and/or prolonged respiratory tract infections in addition to devastating psychological effects.[10]

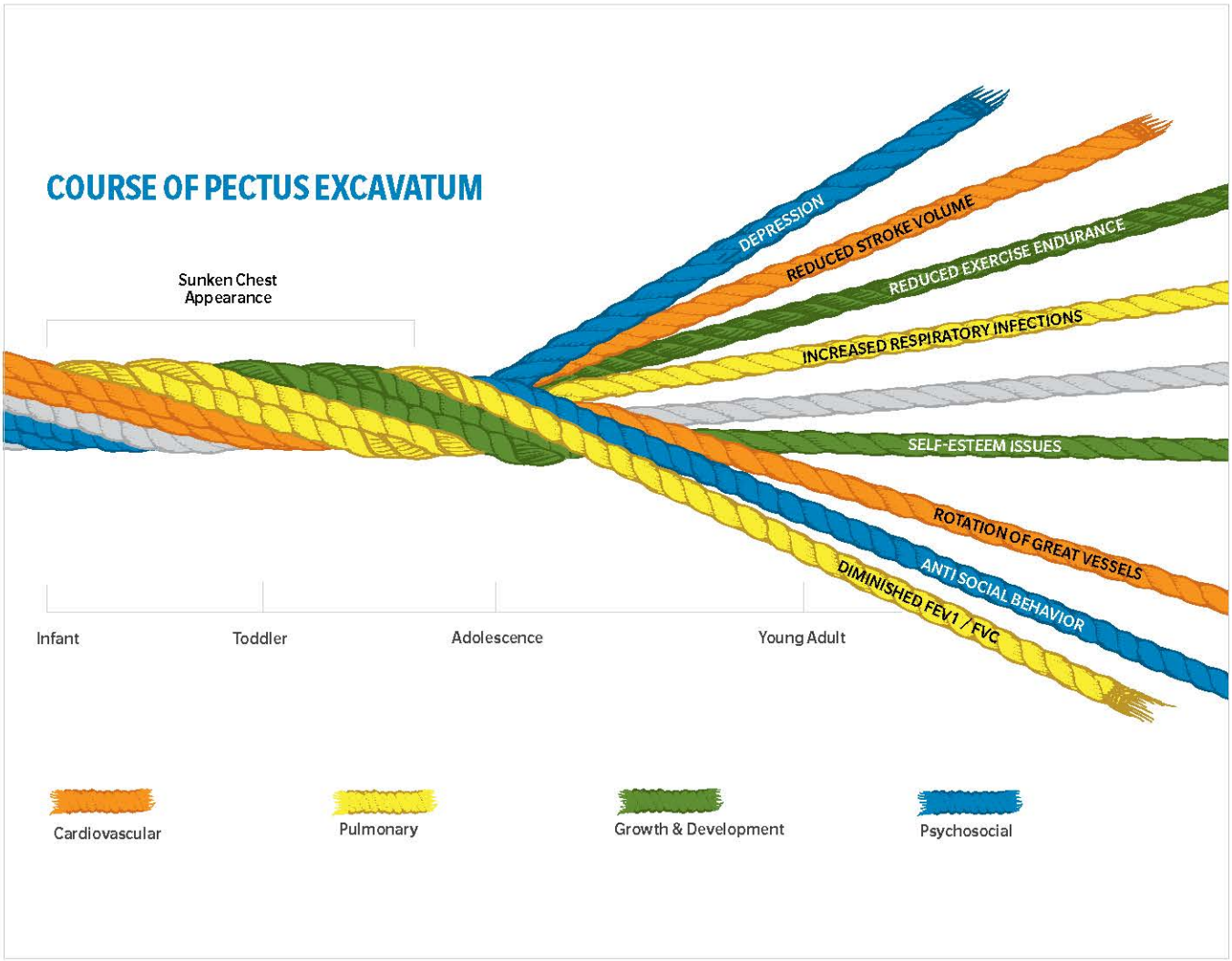

Figure 2: Course of Pectus Excavatum. Pectus excavatum can affect all aspects of life including overall physical and mental well-being, with symptoms progressing as children reach adolescence and young adulthood.

Cardiovascular Impairment

The physical and physiologic impact of PE on the cardiovascular system originates from the posterior displacement of the sternum, resulting in deformity of the myocardium with anterior indentation of the right ventricle, mitral valve prolapse (likely due to deformation of the mitral annulus), and rotation with translocation of the heart into the left hemithorax.[3, 11-13] Patients with PE are suspected of having a reduced stroke volume due to lack of retrosternal space, leading to a diastolic dysfunction in the right ventricle.[14] Other authors have also demonstrated that the rotation of heart, accompanied by torqueing of the great vessels, is an additional contributing factor to the inability to increase stroke volume, leading to a functional restrictive cardiomyopathy that translates into the physical limitation during exertion described in severe cases of PE.[15, 16] An additional study comparing teenagers with pectus excavatum to an age-matched healthy group suggested that PE patients were not capable of increasing their stroke volume to the same extent as healthy peers (Table 1).[17]

Table 1: When compared to healthy controls, teenagers with pectus excavatum exhibit compromised cardiac function in cardiopulmonary exercise tests. Data taken from Lesbo et al., 2011.

	Pectus Patients Mean (95% CI)	Healthy Controls Mean (95% CI)	*P*-value
Minimum heart rate (bpm)	88 (84-91)	86 (81-92)	0.74
Maximum heart rate (bpm)	160 (155-165)	159 (145-173)	0.87
Heart rate increase (bpm)	72 (67-78)	72 (59-85)	0.99
Maximum cardiac index (l/min/m^2)	6.6 (6.3-7.0)	8.0 (7.3-8.8)	0.0001*
Maximum VO_2/kg (ml/min/kg)	26 (24-28)	30 (27-33)	0.04*
Maximum effect (W)	147 (134-159)	159 (142-176)	0.22
Stroke index (ml/beat/m^2)	42 (39-45)	54 (44-64)	0.0022*

*Statistically significant difference between patients and controls. bpm, beats per minute; l/min/m^2, liters per minute per square meter body surface; ml/min/kg, milliliters O_2 per minute per kg body weight.

Pulmonary Function Impairment

The deformation of the rib cage altering the intra-thoracic volume and the capacity of the thorax to effectively expand during the inspiratory effort has been demonstrated by functional studies under exercise conditions, showing more changes than at-rest observations.[18] Other studies have reported values of forced vital capacity (FVC) and forced expiratory volume in 1 second (FEV1) done in healthy volunteers that contrast with lower values of the same respiratory function, measured in pectus patients (Figure 3).[8, 12, 19-26] Oculo-electronic plethysmography can be used to demonstrate that the depressed portion of the chest as seen in severe cases of PE does not move on respiration; while these patients after a corrective surgery, reach a normal static pulmonary function, showing the bell curve of FVC, FEV1, and FEF 25%-75% shifted to higher values by approximately one standard deviation.[27] Although restrictive lung volumes do not invariably improve by repair, enhanced cardiac performance is thought to be partially responsible for improved function.[28]

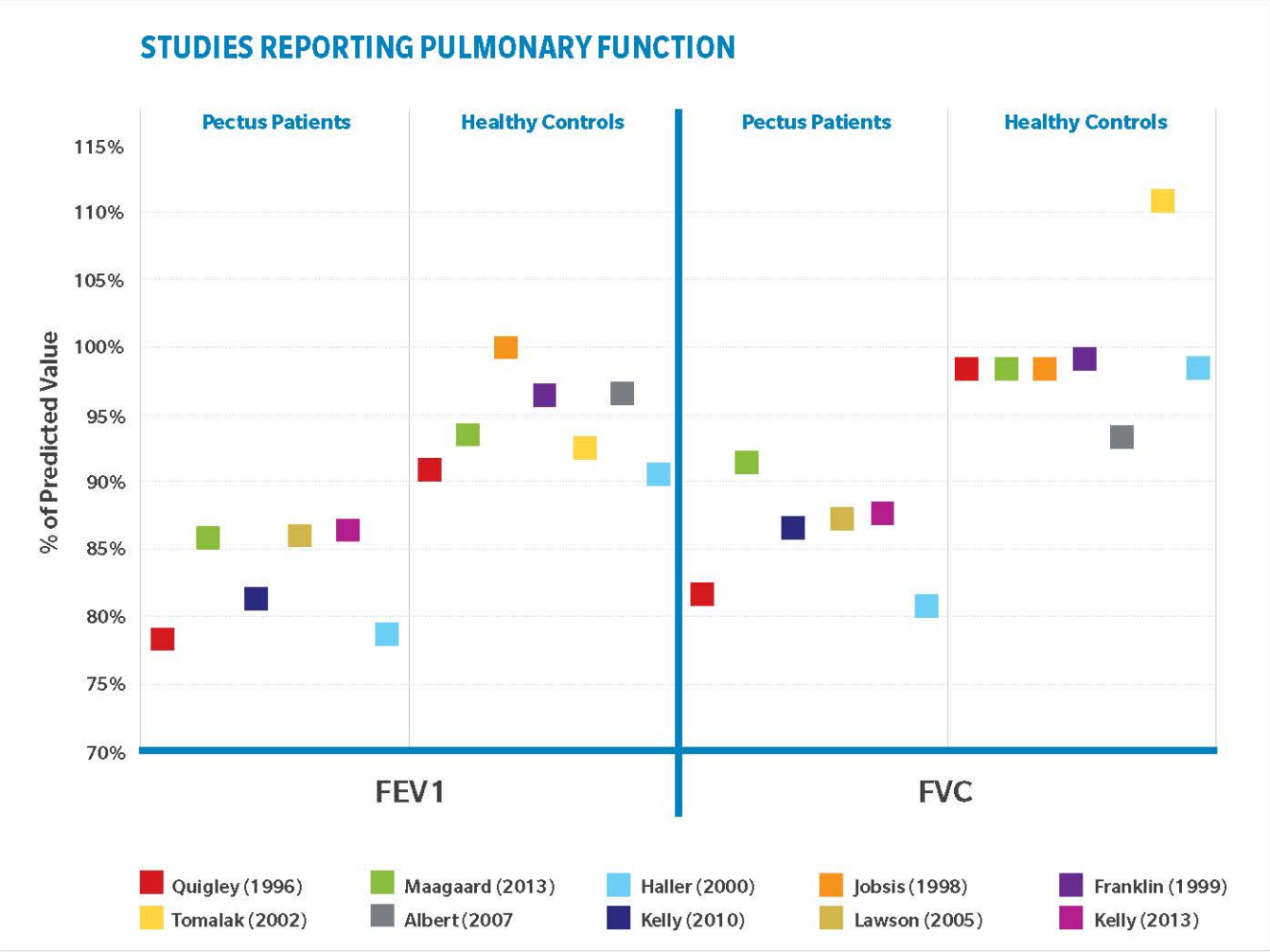

Figure 3: Comparisons of forced expiratory volume in one second (FEV1) and forced vital capacity (FVC) show lower predicted values for pectus patients when compared to studies examining healthy subjects.

Psychosocial Impact

Regardless of the severity of the defect, pectus excavatum often negatively impacts patient quality of life with important effects on mental health and emotional well-being.[4, 5, 29-31] PE can often be the root cause of a negative body image, low self-esteem, somatization, social anxiety and overall dissatisfaction with life. The psychological toll of PE often results in intense feelings of shame leading to avoidance behaviors in social activities, sports, relationships and sexual activities as they progress into puberty and adulthood.[5, 29-31] Results of a study on psychosocial characteristics in PE patients shown in Figure 4 demonstrated how the consequences of PE influence all areas of life, becoming more evident as the adolescence period progresses and difficulties with physical acceptance are more impactful leading to rejection of self-appearance.[31] The reliance on indicators of physical impairment alone discredit the psychological toll of this condition, which can impact quality of life for the patient in ways equal to or exceeding the severity of the physical restrictions.[4, 29-31]

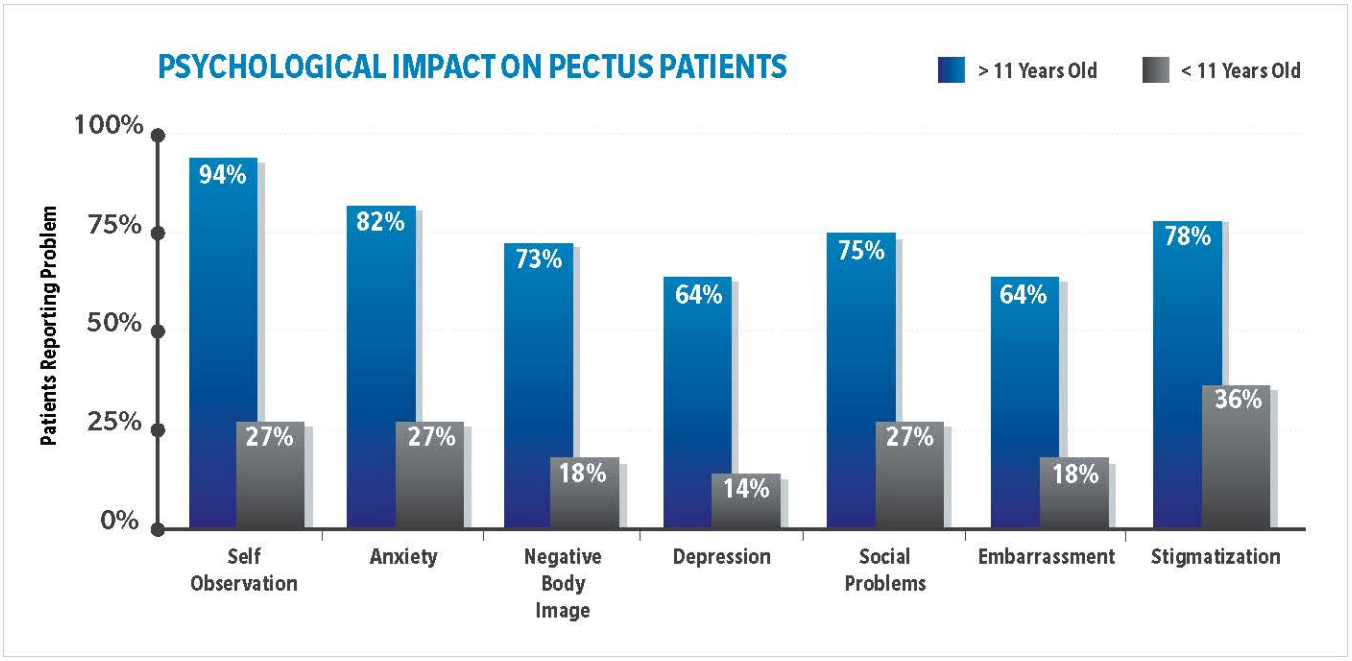

Figure 4: Pectus excavatum is often accompanied by serious psychological effects that worsen as patients age and can exceed the impact of physical restrictions. Data taken from Einsiedel et al., 1999.

Human Growth and Development

Adequate physical activity in each developmental stage is thought to have a positive impact on human mental and psychosocial health, conditioning their physical, cognitive, and academic performance. While the extent to which anteroposterior chest compression caused by pectus excavatum may limit exercise capacity remains controversial, patients with severe PE, particularly adolescents, have shown limitations on endurance fitness and cardiorespiratory functional reserve.[32] Previous studies have provided compelling evidence of a variety of ventilatory deficiencies in patients with pectus deformities that might prove limiting to exercise performance.[20, 32-36]

Minimally Invasive Repair: The Nuss Procedure

For defects that extend beyond a mild cosmetic disorder, untreated PE results in progressive cardiopulmonary consequences for patients.[37] Minimally invasive repair of pectus excavatum (MIRPE), also known as the Nuss procedure, is the first procedure that corrects pectus excavatum without an open incision into the pulmonary space.[38] Figure 5 shows a brief overview of the Nuss procedure, which involves inserting a U-shaped metal bar through small incisions on either side of the rib cage (A) and positioning the bar (B) so that it pushes the anterior chest wall outward (C). One or more bars are inserted and secured to the lateral chest wall with stabilizing plates, stretching the chest wall without the need for tissue removal or dissection. Bars remain implanted for 2-4 years, allowing the chest wall time to remodel before removal.[39] Since its inception in the 1990's, the Nuss procedure has become the standard of care for pectus excavatum repair and has an established track record of successful defect repair with improvement in cardiovascular function, pulmonary function, exercise tolerance, and patient quality of life.[4,38]

Figure 5: The Nuss procedure involves inserting a U-shaped metal bar (A) through small incisions on either side of the rib cage and then flipping the bar (B), correcting the defect without the need for tissue removal or dissection (C).

Cardiovascular Improvement

Pectus excavatum can significantly impair cardiac function by producing cardiac displacement and/or compression (Figure 6).[1-3] A number of cardiac metrics have shown improvement after the Nuss procedure. For example, a study in teenagers with pectus excavatum (average age: 15 years old) experienced significant increases in both diastolic and systolic diameters over baseline at the time of bar removal 3 years after the initial Nuss procedure ($p \leq 0.001$). Both the right and left ventricle had increased diastolic diameters (L: 4.9 cm vs 4.6 cm, p=0.0002; R: 2.3 cm vs 2.1 cm, p=0.001), while only the left ventricle showed increased systolic diameter (3.3 cm vs 3.0 cm, p=0.0001).[20] Two other studies provide complementary data on cardiac benefits of Nuss repair. In one, stroke volume and cardiac output both increased significantly at 3 years post procedure compared with preoperative values. The mean stroke volume increased to 84 mL/beat/m^2 from 69 mL/beat/m^2 pre-operatively ($p<0.05$), and mean cardiac output reached 5.4 mL/min/m^2 from 4.7 mL/min/m^2 before repair.[34] Another study showed trends toward increased stroke volume (75 mL/beat/m^2 vs 68 mL/beat/m^2, p=0.05) and cardiac output (5.1 mL/min/m^2 vs 4.7 mL/min/m^2, p=0.07).[35]

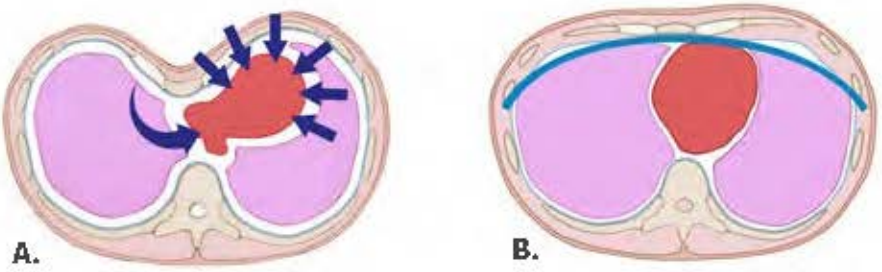

Figure 6. Pectus excavatum can significantly impair cardiac function by producing cardiac displacement and/or compression and torsion of the great vessels (A). Minimally invasive repair of the defect allows the heart and great vessels to return to a normal position and shape (B).

Pulmonary Function Improvement

Many studies have noted benefits to pulmonary function after Nuss repair of pectus excavatum. Three of four separate pediatric populations experienced significant increases in FEV1 measurements following bar removal after the Nuss procedure (Figure 7).[20,34-36] The baseline values for the four studies varied significantly (63% – 91%), but all populations experienced a mean increase in FEV1 of at least 5%.[20, 34-36, 40] The study with the greatest restriction in FEV1 also showed the largest benefit from Nuss repair — increasing from 63% to 85%.[36] Outcomes in similar populations measured after Nuss repair but before the bars were removed resulted in more subdued outcomes, suggesting that full pulmonary function benefits are not realized while the bar remains implanted.[7,40,41]

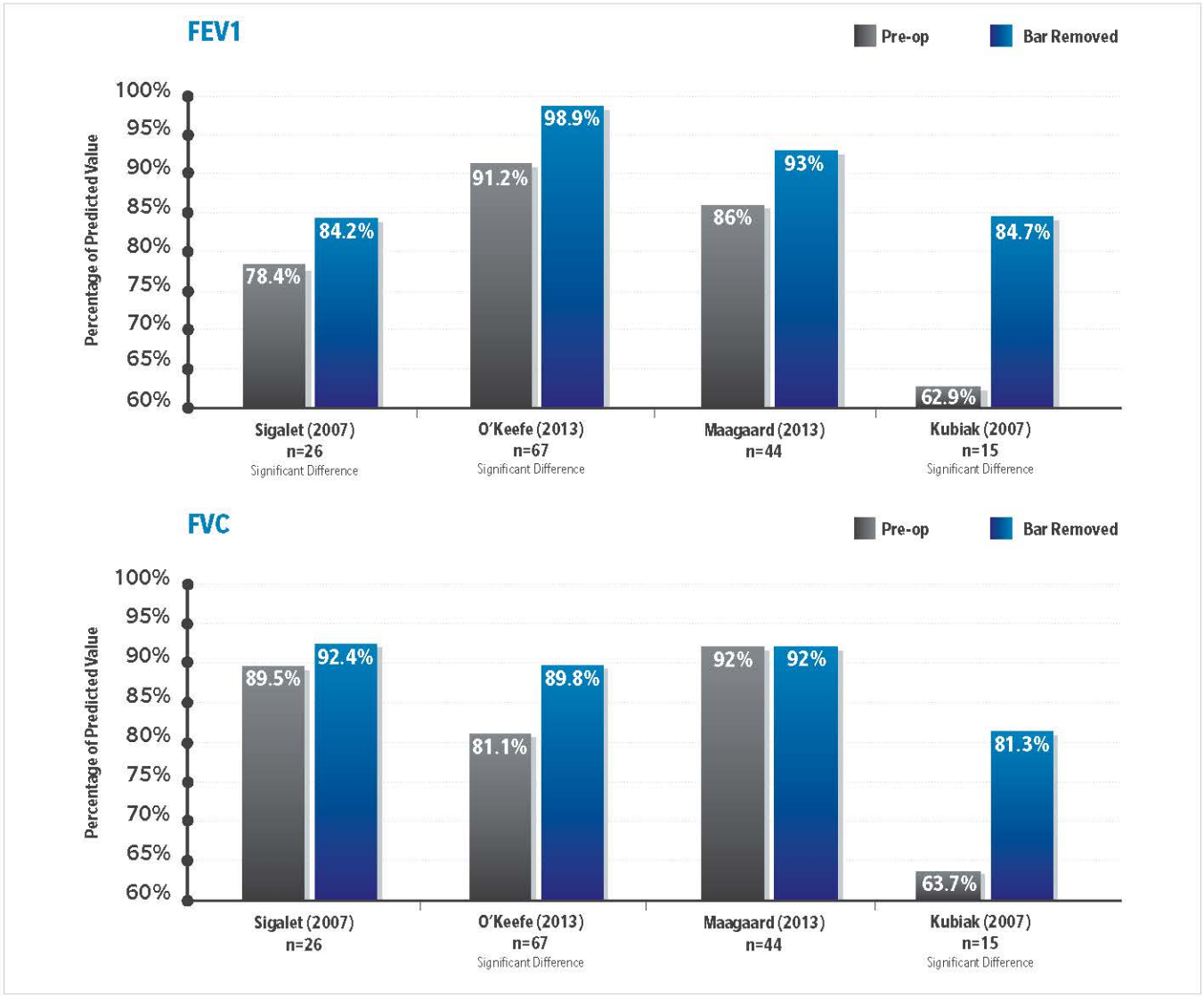

Figure 7. Comparisons of forced expiratory volume in one second (FEV1) and forced vital capacity (FVC) show improvements in pulmonary function after correction of pectus defect with the Nuss procedure when compared to preoperative values.

Cardiopulmonary Function Improvement During Exercise

The importance of physical activity in children and adolescents' growth and development is not disputed.[42] Adequate physical activity in each developmental stage is thought to have a positive impact on human mental and psychosocial health, conditioning their physical, cognitive, and academic performance. Patients' ability to participate in vigorous exercise is one of the most distressing problems with pectus excavatum, often with lifestyle altering consequences.[20] Failure to engage in vigorous- or moderate-intensity physical activity contributes to a decrease in flexibility, muscular strength, diminished cardiorespiratory capacity and a greater risk for other diseases.[43]

Prior to undergoing the Nuss procedure, a population of teenagers with pectus excavatum exhibited significant differences in maximum cardiac index and stroke index compared with healthy controls (Figure 8). Before correction, the pectus cohort had a maximum cardiac index of 6.6 mL/min/m^2. Patients saw increases in cardiac index at 1 year and at 3 years (coinciding with bar removal). At 1 year, the maximum cardiac index for the pectus cohort already showed significant differences compared with baseline (mean value: 7.3 mL/min/m^2; $p<0.001$) and at 3 years, the mean value showed no difference when compared with that of the healthy controls (8.1 mL/min/m^2 vs 8.3 mL/min/m^2, $p=0.57$). The stroke index showed similar results: baseline values for the pectus cohort averaged 42 mL/beat/m^2 (compared with 54 mL/beat/m^2 in the healthy controls), which had increased to 44 mL/beat/m^2 after 1 year. At bar removal (3 years post procedure), the mean stroke index for the pectus cohort showed significant improvement over baseline ($p<0.001$) and was indistinguishable from that of healthy controls (50 mL/beat/m^2 vs 50 mL/beat/m^2, $p=0.993$). Increases in maximum oxygen uptake after repair that resolved to values similar to those in the control group at 3 years after Nuss repair were also noted (29 mL/min/kg vs 31 mL/min/kg, $p=0.43$).[20]

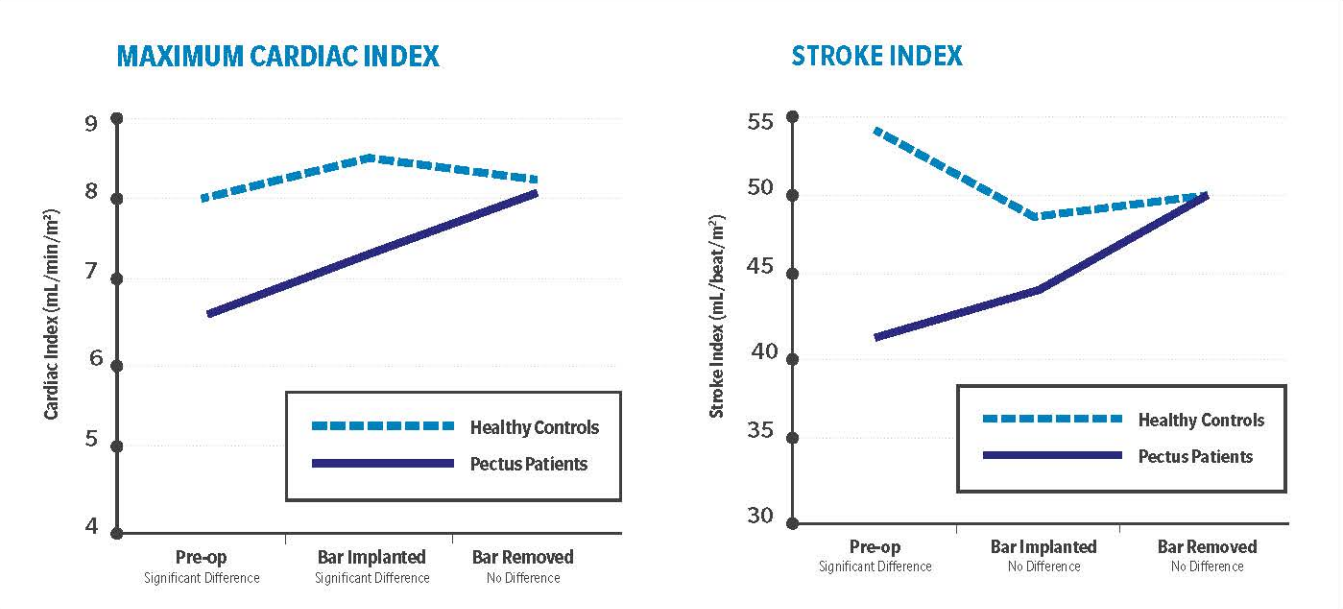

Figure 8. Cardiopulmonary function in pectus patients improved after undergoing the Nuss procedure and continued to improve to levels similar to healthy controls after removal of the bars. Data taken from Maagaard et al., 2013.

Quality of Life Improvement

The majority of studies in patients with pectus excavatum focus on functional measures of improvement. While these metrics are standard outcomes for showing the physical impact of repair on patients' daily function, this singular focus on physical data disregards the complementary part of the equation: how the condition impacts patients in a holistic manner, particularly their psychologic state and quality of life.

In one study focused on quality of life measures, both patients and their parent or caregiver figure reported high satisfaction scores with the results of the Nuss procedure (4.6 out of 5, Figure 9). In addition to positive changes that patients reported surrounding self-perception and body image after the corrective surgery, their caregivers/parents appeared to concur with 82% estimating that the patient was either "satisfied" or "very satisfied" with the result of the operation. Independent of this result, significant improvements in various scales were observed. The Global Severity Index, a test designed to help quantify a patient's severity-of-illness reflecting both the number of symptoms and intensity of perceived distress, shows significant improvement ($P = 0.03$) in repaired pectus patients. Results demonstrate the improvement of the psychological distress in these patients.[44] Other studies have found similar results, suggesting that Nuss repair significantly increases patients' satisfaction with their appearance compared with pre-repair baseline values.[29-31] Improvements in both physical and psychosocial measures of health-related quality of life support the idea that Nuss repair of pectus excavatum may have significant benefits to both physical and psychologic effects of this condition.[45]

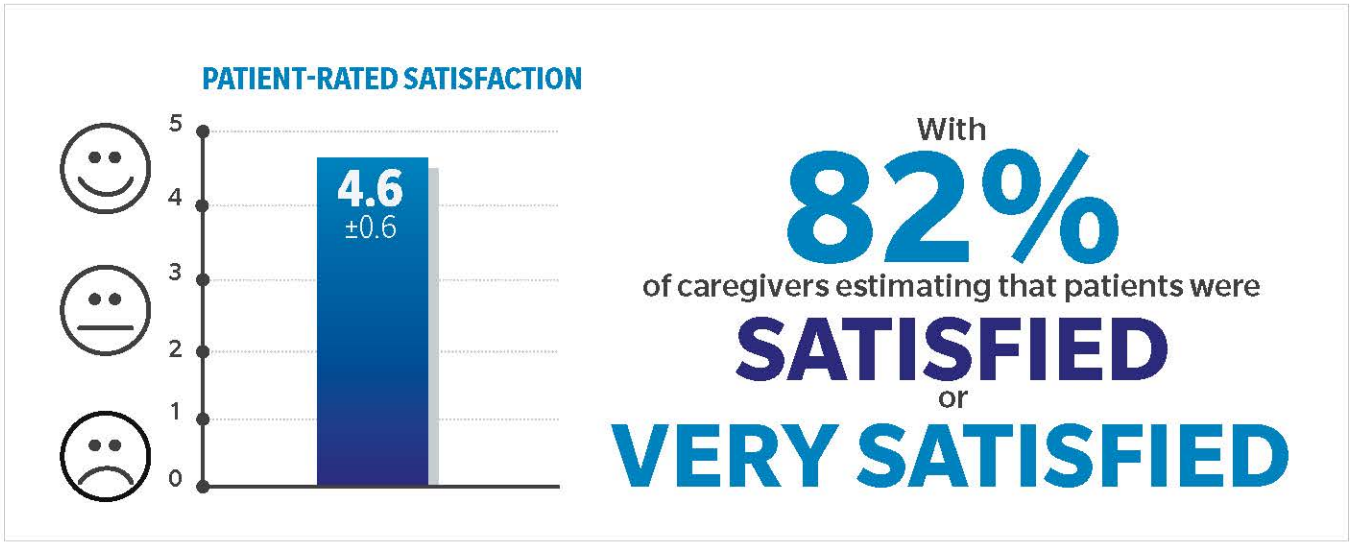

Figure 9. Patient-rated satisfaction of the Nuss procedure was very high and 82% of parents and caregivers estimated that patients were either "satisfied" or "very satisfied" with the result of the operation. Data taken from Hadolt et al., 2011.

Conclusions

This diverse array of physical and psychologic improvements after Nuss repair of pectus excavatum provides evidence that may help revise long-standing beliefs that surgical repair provides only cosmetic enhancement, leading to more proactive management of this significant condition and ultimately better outcomes for patients. While the ill effects of this congenital malformation are manifested mainly in late adolescence or young adulthood, the development of cardiovascular, pulmonary, and psychosocial effects associated with PE start in childhood and early adolescence. Surgical correction of the defect has been proven to improve physical and psychosocial outcomes and promote health-enhancing behaviors and positive long-term effects associated to physical activity.[26, 29]

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