Clinical Commissioning Policy: Surgery for pectus deformity (all ages)

NHS England Reference: 170113P

NHS England INFORMATION READER BOX		
Directorate		
Medical	Operations and Information	**Specialised Commissioning**
Nursing	Trans. & Corp. Ops.	Commissioning Strategy
Finance		

Publications Gateway Reference:	**07603**
Document Purpose	Policy
Document Name	Surgery for pectus deformity (all ages)
Author	Specialised Commissioning Team
Publication Date	22 February 2019
Target Audience	CCG Clinical Leaders, Care Trust CEs, Foundation Trust CEs , Medical Directors, Directors of PH, Directors of Nursing, NHS England Regional Directors, NHS England Directors of Commissioning Operations, Directors of Finance, NHS Trust CEs
Additional Circulation List	
Description	Not for routine commissioning
Cross Reference	
Superseded Docs (if applicable)	
Action Required	
Timing / Deadlines (if applicable)	
Contact Details for further information	england.specialisedcommissioning@nhs.net

Document Status

This is a controlled document. Whilst this document may be printed, the electronic version posted on the intranet is the controlled copy. Any printed copies of this document are not controlled. As a controlled document, this document should not be saved onto local or network drives but should always be accessed from the intranet.

Clinical Commissioning Policy: Surgery for pectus deformity

First published: February 2019

Prepared by NHS England Specialised Services Clinical Reference Group for Radiotherapy and Specialised Cancer Surgery

Published by NHS England, in electronic format only.

Policy Statement

NHS England will not routinely commission surgery for pectus deformity in accordance with the criteria outlined in this document.

In creating this policy NHS England has reviewed this clinical condition and the options for its treatment. It has considered the place of this treatment in current clinical practice, whether scientific research has shown the treatment to be of benefit to patients, (including how any benefit is balanced against possible risks) and whether its use represents the best use of NHS resources.

Equality Statement

Promoting equality and addressing health inequalities are at the heart of NHS England's values. Throughout the development of the policies and processes cited in this document, we have:

- given due regard to the need to eliminate discrimination, harassment and victimisation, to advance equality of opportunity, and to foster good relations between people who share a relevant protected characteristic (as cited under the Equality Act 2010) and those who do not share it; and
- given regard to the need to reduce inequalities between patients in access to, and outcomes from healthcare services and to ensure services are provided in an integrated way where this might reduce health inequalities.

Plain Language Summary

About Pectus deformity

Pectus deformity is a term used to describe a group of conditions associated with the malformation of the chest wall. In most cases, a pectus deformity will be present at birth, however, it will usually only become obvious and visible during early adolescence, when growth is rapid. Diagnosis is made by physical examination.

There are two main types of pectus deformity:

- Pectus excavatum, in which the breastbone is sunken inwards and the chest looks hollow. Sometimes this is called “funnel chest”. This is the most common type of deformity; and
- Pectus carinatum, in which the breastbone is raised and the chest is pushed out. Sometimes this is called “pigeon chest”.

Both conditions are more likely to affect males than females (Krasopoulos et al 2010, Goresky et al 2004, Kelly et al 2004) and both may be inherited, often being present in several members of the same family and associated with other congenital diseases including scoliosis of the spine, Marfan’s syndrome and Ehlers-Danlos syndrome (Goretsky et al 2004).
The impact of a pectus deformity can vary substantially, ranging from mild and symptomless to severe and impacting on both lung (pulmonary) and heart (cardiac) function. Psychologically, the deformity usually has little impact, but in some cases it can cause psychological distress leading to concerns about appearance, withdrawal and social isolation.

About current treatments

There are a range of surgical and non-surgical treatments available to manage the condition and individuals with a pectus deformity may be referred to a thoracic surgical clinic for advice. Treatment options are determined by assessment of the type of pectus deformity, degree of deformity, simple versus mixed deformity, and determination of whether the deformity is isolated or part of a syndrome.

In most cases, while surgery can correct the chest wall deformity, surgical intervention does not take place. This is because the majority of people experience only mild physical or psychological symptoms associated with having a pectus deformity. In these cases, non-surgical options include posture, exercise programmes, bracing and psychological support.

Where surgical intervention does take place, there are two types of procedure available:

- The Nuss procedure, which is a minimally invasive intervention generally only used to treat cases of pectus excavatum. It involves placing one or two steel bars under the breastbone with the aim of raising it and correcting the abnormal shape. Each bar, bent into a curve to fit the patient's chest, is inserted through small openings in the chest. The bar (or bars) is/are usually removed within a few years of placement; and
- The Ravitch procedure, which can be used to treat both pectus excavatum and pectus carinatum. In this technique the rib cartilages are cut away on each side and the sternum is flattened so that it will lie flat. One or more permanent bars or struts are inserted to ensure the sternum keeps its new shape.

This policy examines the continued place of surgical intervention in the management of pectus deformity.

What we have decided

NHS England has carefully reviewed the evidence for the surgical correction of pectus deformities. We have concluded that there is not enough evidence to routinely commission the intervention.

1 Introduction

Clinical indication

Pectus abnormalities cover a range of deformities affecting the anterior chest wall, specifically the sternum and adjacent rib cartilages. Pectus excavatum (PE) and pectus carinatum (PC) are the most common of these. PE, or 'funnel chest', appears as a depression of the sternum; it may be asymmetrical, with the right side deeper than the left (de Oliveira Carvalho, 2014). It can be present at birth or may develop during childhood and adolescence. PC, also known as 'pigeon chest', is caused by the sternum pushing out so the middle of the chest is more pronounced (Goretsky et al 2004). It usually manifests at the time of a growth spurt in the early teenage years (Fokin et al 2009). If not corrected, the deformity is permanent.

Both deformities may be inherited, often being present in several members of the same family and associated with other congenital diseases including scoliosis of the spine, Marfan's syndrome and Ehlers-Danlos syndrome (Goretsky et al 2004).

Pectus deformities vary from mild and asymptomatic to a more severe form which can alter measures of pulmonary and cardiac function. Diagnosis is by physical examination (Lain et al 2017). Psychologically, the deformity usually has little impact, but in some cases it can cause significant psychological distress leading to concerns about appearance, withdrawal and social isolation (Steinman et al 2011).

There are a number of non-surgical management options to support people diagnosed with a pectus deformity, including posture and exercise programmes, bracing and psychological support.

Proposed intervention

There are two types of surgical procedure available for the correction of pectus deformity:

- Nuss procedure, which is a minimally invasive procedure generally only used to treat cases of PE; and
- Ravitch procedure, which is an open surgical procedure that can be used to treat both cases of PE and PC.

The Nuss procedure involves placing one or two steel bars under the breastbone with the aim of raising it and correcting the abnormal shape. Each bar, bent into a curve to fit the patient's chest, is inserted through small openings in the chest. The bar (or bars) is usually removed within a few years of placement.

In the modified Ravitch procedure, the rib cartilages are cut away on each side and the sternum is flattened so that it will lie flat. One or more permanent bars or struts are inserted to ensure the sternum keeps its shape.

Treatment choice is determined by assessment of the type of pectus deformity, degree of deformity, simple versus mixed deformity, and determination of whether the deformity is isolated or part of a syndrome. The severity of the deformity can be assessed radiologically using the Haller index, defined as the lateral external distance divided by the distance between the external deepest point of the chest and external spinous process.

2 Definitions

Bracing – the use of a back brace to stop the deformity getting worse. It is used in children, often until they stop growing.

Ehlers-Danlos syndrome – is a group of inherited disorders that affect the connective tissues, primarily skin, joints and blood vessel walls.

Marfan's syndrome - is a genetic disorder that affects the body's connective tissue.

Nuss – is a minimally invasive procedure to repair pectus excavatum.

Pectus Carinatum – is where the sternum is raised so the chest appears pushed out.

Pectus Excavatum – is where the sternum is depressed and the chest looks hollow.

Ravitch – is an open surgical procedure for the treatment of pectus excavatum and pectus carinatum.

Scoliosis - is a condition in which the spine can be curved or twisted.

3 Aims and Objectives

This policy considered: surgical correction of pectus deformity.

The objectives were to: establish, via an evidence review, whether:

- Surgical correction improves cardiorespiratory reserve, functional and physical outcomes;
- Surgical intervention significantly improves the psychological well-being and quality of life for individuals with pectus deformities;
- There is a relationship between the degree of the pectus deformity (as quantified by the Haller index or other objective assessment) and the level of psychological distress experienced;
- There are subgroups in which surgery produces a greater improvement in mental wellbeing than others;
- Surgical volume impacts on the outcome of surgery (infection and revision rates);
- There is evidence with regard to quality, safety, and adverse events associated with surgical correction; and
- There is an evidence base for eligibility criteria and thresholds for surgery.

4 Epidemiology and Needs Assessment

Birth incidence of pectus deformity is estimated to be between 1 in 400 and 1 in 1000, of which 87% will be PE, 5% PC and the remainder a combination of the two or other very rare chest deformities (Lomholt et al 2016, Krasopoulos et al 2010, Goresky et al 2004). Both PE and PC are more likely to present in males, with a male to female ratio of between 3:1 and 9:1 (Krasopoulos et al 2010, Goresky et al 2004, Kelly et al 2004).

NHS activity data indicates that 250 surgical procedures to correct pectus deformity were carried out in England during 2017/18.

5 Evidence Base

NHS England has concluded that there is not sufficient evidence to support the routine commissioning of surgical treatment for pectus deformity

Psychological, Social and Behavioural Outcomes

Twenty six published studies were identified evaluating the effectiveness of corrective pectus surgery relating to improve psychological, social and behavioural outcomes. There were a number of serious weaknesses identified in the evidence that was found. The studies were not well-controlled. Four of the studies used a before-and-after design with no patient control group, which made it impossible to discern whether any changes observed are because of the intervention or because of another factor, such as the passage of time or some other change in participants' circumstances. Having unoperated patients with higher pre-operative body satisfaction as controls is clearly inappropriate because the reported differences are likely to reflect disease severity rather than the effect of surgery. The use of healthy controls provides no information on the difference that surgery may make to those with pectus deformity.

Although the studies all reported statistically significant results, they provide little information on the functional significance to patients of the differences that they found. A change may be large enough to be statistically significant, i.e., to make chance an unlikely explanation, but too small to be of real value to the patient. In many cases, patients' and parents' scores did not indicate the existence of major psychosocial or other difficulties before surgery, limiting the improvement that surgery could provide.

Outcome measures varied between studies and were often opaque. The terminology used in the technical reporting of psychometric questionnaires is often not suitable for direct translation into clinical commissioning policy. The studies did not make clear what each outcome measure meant and what functional effect a change might have in a patient's life. It was difficult to assess the relevance and importance of the

differences reported. The studies lacked any objective measures of effect on social integration, such as school attendance or participation in sport.

There were many important data the authors did not report, such as whether participants had undergone other treatment before or after surgery, the extent to which activities of daily living were prevented by their deformity and how often the procedure had adverse effects.

Cardiorespiratory reserve, functional and physical outcomes

Relevant research studies highlighted by the evidence review of surgical intervention to correct pectus deformity are largely either limited to case series and reports or studies that were not well controlled. The meta-analysis did not attempt to address statistical heterogeneity between studies or take into account surgical skill variations amongst individual surgeons, between centres and over time. The absence of a standardised measure/scale to weigh clinical benefits (physical, psychological and quality of life) against the significant morbidity caused by the procedures presents a challenge to any conclusion regarding benefits of intervention.

Johnson et al, 2014 found no linkage between ages of operative treatment with outcomes. There was no clear difference in outcomes between the Nuss and Ravitch populations across all age groups, but slightly better outcomes in the Nuss paediatric group as compared to all other groups. Nasr et al, 2010 found no difference in patient satisfaction between both techniques among studies looking at this outcome. A meta-analysis of 2476 cases (1555 Nuss, 921 open surgery) from 23 international studies (Chen et al, 2012) reported significant overall improvement in lung function 3 years after surgery, measured as separate net changes in forced expiratory volume over 1 second (FEV1), forced vital capacity (FVC), vital capacity (VC), and total lung capacity (TLC). The improvement was reported to be better with the Nuss procedure compared to open surgery. The preoperative pulmonary function tests lay at the lower end of the normal range and again there is no report of the functional status at the pre- or post-operative stages. This together with the absence of a non-intervention or healthy comparator arm in any of the studies, means that these findings cannot be used to draw an inference on the clinical effectiveness of pectus procedures on lung function. Cardiovascular function after surgery was reported to

be improved by greater than one-half standard deviation. However, no supporting analysis was included in the publication. Other large case series (Kelly et al, 2013. Žganjer et al, 2011) report positive improvement of chest wall in varying degrees as well as improvement in pulmonary function. Most studies report 80-90% good to excellent anatomic surgical outcomes. Given the limitations in the study design, the overall evidence in this category needs to be viewed with caution.

There were no studies that directly compared the impact of surgeon volume and outcomes of surgery. In a retrospective review of all primary Nuss procedure repairs of pectus excavatum performed in a one large US centre over 21 years, complications decreased markedly over 21 years since surgery was first offered in the centre. Bar displacement rate requiring surgical repositioning decreased from 12% in the first decade to 1% in the second decade (Kelly et al, 2010). This provides a limited view of the impact of surgical experience and patient volume on outcomes.

The literature review did not find any randomised control trials or high quality meta-analysis that could further update the comparative efficacy of different types of surgery or provide a comparison with a no-intervention group. The best available evidence comes from a systematic review of 39 studies involving 807 adult and 2716 paediatric cases (Johnson et al, 2014) which focused on comparison of the Ravitch, Nuss, and other surgical treatments for pectus excavatum across age groups. The analysis showed that complication rates varied across studies, however, Nuss and Ravitch procedures were generally safe for paediatric and adult patients with no perioperative mortality reported. Re-operation rates in adults were highest for implant procedures at 18.8% followed by Nuss 5.3% and Ravitch 3.3% but there was no significant difference in re-operation rates in children. Nasr et al, 2010 found that there was no significant difference in overall complication rates between both techniques in the nine studies included in the meta-analysis. Looking at specific complications, postoperative pneumothorax and haemothorax, the rate of reoperation because of bar migration or persistent deformity was significantly higher in the Nuss group. Most case series identified major and minor complications related with the surgery ranging from allergy to nickel (Nuss bars), pneumothorax, haemothorax and pericardial tears in perioperative period to bar displacement and asymmetrical corrections that required re-operations.

No evidence was found about the relationship between the degree of the pectus deformity and change in psychological outcomes, about sub-groups in which surgery produces a greater improvement in mental wellbeing and whether the incorporation of psychosocial therapies into surgical management affects outcome

Conclusion

The evidence that was found is not sufficient to conclude that the physical psychological, social and behavioural benefits of surgical treatment of pectus deformities are sufficient to justify its use.

6 Documents which have informed this Policy

The documents that have informed this policy are:

- NHS England 2015. Evidence Review: Surgical correction for Pectus deformity (all ages) Public consultation document. NHS England;
- NHS England 2018. Evidence Review: Surgical correction for Pectus deformity (all ages) Public consultation document. NHS England;
- NICE 2009. Placement of pectus bar for pectus excavatum (also known as MIRPE or the Nuss procedure). https://www.nice.org.uk/Guidance/ipg310

7 Date of Review

This document will be reviewed when information is received which indicates that the policy requires revision.

References

Bahadir AT, Kuru Bektasoglu P, Cakiroglu Eser A, Afacan C, Yuksel M. 2017. Psychosocial functioning in pediatric patients with pectus excavatum and pectus carinatum. Turkish Journal of Medical Sciences. 2017;47(3):771-7.

de Oliveira Carvalho PE, da Silva MVM, Rodrigues OR, Cataneo AJM 2014.Surgical interventions for treating pectus excavatum. Cochrane Database of Systematic Reviews 2014, Issue 10. Art. No.: CD008889. DOI:1002/14651858.CD008889.pub2.

Fokin AA, Steuerwald NM, Ahrens WA, Allen KE. 2009. Anatomical, histologic, and genetic characteristics of congenital chest wall deformities. Semin Thorac Cardiovasc Surg; 21: 44-57.

Goretsky MJ, Kelly RE, Croitoru D, Nuss D.2004. Chest wall anomalies. Pectus excavatum and pectus carinatum AdolescMed. 2004;15(3):455–71.

Hadolt B, Wallisch A, Egger JW, Hollwarth ME. 2011 .Body-image, self-concept and mental exposure in patients with pectus excavatum. Pediatric Surgery International. June;27(6):665-70.

Jacobsen EB, Thastum M, Jeppesen JH, Pilegaard HK. 2010. Health-related quality of life in children and adolescents undergoing surgery for pectus excavatum. European Journal of Pediatric Surgery.;20(2):85-91.

Kelly RE, Jr., Cash TF, Shamberger RC, Mitchell KK, Mellins RB, Lawson ML, et al. 2008. Surgical repair of pectus excavatum markedly improves body image and perceived ability for physical activity: Multicenter study. Pediatrics. Dec 2008

Krasopoulos G, Goldstraw P. 2011 Minimally invasive repair of pectus excavatum deformity. European Journal of Cardio-thoracic Surgery. February;39(2):149-58.

Kuru P, Bostanci K, Ermerak NO, Bahadir AT, Afacan C, Yuksel M. 2015 Quality of life improves after minimally invasive repair of pectus excavatum. Asian Cardiovascular and Thoracic Annals. 17 Mar;23(3):302-7.

Lain A, Garcia L, Gine C, Tiffet O and Lopez M 2017. New Methods for Imaging Evaluation of Chest Wall Deformities. Front. Pediatr. 5:257. doi: 10.3389/fped.2017.00257

Lawson ML, Cash TF, Akers R, Vasser E, Burke B, Tabangin M, Welch C, Croitoru DP, Goretsky MJ, Nuss D, Kelly RE Jr 2003. A pilot study of the impact of surgical repair on disease-specific quality of life among patients with pectus excavatum. J Pediatr Surg 38: 916-8.

Lomholt JJ, Jacobsen EB, Thastum M, Pilegaard H. 2016 A prospective study on quality of life in youths after pectus excavatum correction. Ann Cardiothorac Surg. Sep;5(5):456-65.

Luo L, Xu B, Wang X, Tan B, Zhao J. 2017. Intervention of the Nuss Procedure on the Mental Health of Pectus Excavatum Patients. Ann Thorac Cardiovasc Surg. Aug 20;23(4):175-80.

Moss TP. 2005. The relationships between objective and subjective ratings of disfigurement severity, and psychological adjustment. Body Image.2(2):151–159

NHS England 2015. Evidence Review: Surgical correction for Pectus deformity (all ages) Public consultation document. NHS England.

Steinmann C, Krille S, Mueller A, Weber P, Reingruber B, Martin A, 2011. Pectus excavatum and pectus carinatum patients suffer from lower quality of life and impaired body image: A control group comparison of psychological characteristics

prior to surgical correction. European Journal of Cardio-thoracic Surgery. November;40(5):1138-45.

Kelly, Robert E.; Shamberger, Robert C.; Mellins, Robert B.; Mitchell, Karen K.; Lawson, M. Louise; Oldham, Keith; Azizkhan, Richard G.; Hebra, Andre V.; Nuss, Donald; Goretsky, Michael J.; Sharp, Ronald J.; Holcomb, George W.; Shim, Walton K. T.; Megison, Stephen M.; Moss, R. Lawrence; Fecteau, Annie H.; Colombani, Paul M.; Bagley, Traci C.; Moskowitz, Alan B.. Prospective multicenter study of surgical correction of pectus excavatum: design, perioperative complications, pain, and baseline pulmonary function facilitated by internet-based data collection. J. Am. Coll. Surg. 2007;16(6):639-642.b.

Coelho, Marlos de Souza; Silva, Ruy Fernando Kuenzer Caetano; Bergonse Neto, Nelson; Stori, Wilson de Souza; dos Santos, Anna Flávia Ribeiro; Mendes, Rafael Garbelotto; Fernandes, Lucas de Matos. Pectus excavatum surgery: sternochondroplasty versus Nuss procedure. Ann. Thorac. Surg. 2009;44(5):888-892.c.

Loff, Steffan; Sauter, Hartwig; Wirth, Thomas; Otte, Ralf. Highly Efficient Conservative Treatment of Pectus Carinatum in Compliant Patients. Eur J Pediatr Surg 2015;96(1):272-278.d.

Steinmann, Cornelia; Krille, Stefanie; Mueller, Astrid; Weber, Peter; Reingruber, Bertram; Martin, Alexandra. Pectus excavatum and pectus carinatum patients suffer from lower quality of life and impaired body image: a control group comparison of psychological characteristics prior to surgical correction. Eur J Cardiothorac Surg 2011;25(5):421-424.