American Pediatric Surgical Association

Pectus Carinatum Guideline

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Synopsis

This clinical practice guideline serves as a collection of recommendations to assist pediatric surgeons and pediatricians in the evaluation and management of children with pectus carinatum. The recommendations resulted from an integration of a comprehensive systematic review of the medical literature and expert opinion regarding pectus carinatum. As a guideline these recommendations should not supplant the clinical judgment of the treating physician and should not be applied as a general protocol for every child with pectus carinatum.

Definitions

Pectus carinatum is defined as a continuum of anterior chest wall deformities noted by the protrusion of the sternum and associated convex deformity of the adjacent costal cartilages. The most common categorizations are chondrogladiolar or chondromanubrial subtypes. Chondrogladiolar pectus carinatum consists of protrusion of the costal cartilages with deformation of the gladiolus segment, or so-called body of the sternum, which may be symmetric or asymmetric in its configuration. The rarer chondromanubrial subtype of pectus carinatum represents protrusion of the manubrium of the sternum and the adjacent costal cartilages. [1, 2]

Clinical Presentation

Pectus carinatum occurs with a male predominant gender ratio of 4:1.[3] Although it can present in early childhood, it often is not apparent until puberty at which time it can progress in severity during rapid linear growth. Pectus carinatum may present with or without symptoms. The most common symptoms elicited may include exercise intolerance, chest pain, chest wall tenderness, shortness of breath, palpitations, or wheezing.[4, 5, 6, 7, 8] In children whose chest wall significantly deviates from normal, reconstructive and psychosocial concerns are reasons families may seek medical attention for pectus carinatum.[9] An association with scoliosis is most common, but rarer associations with congenital heart disease, Marfan's syndrome, and other connective tissue disorders have been reported.[10, 11, 12, 13, 14] In adults, who did not have chest wall reconstruction in childhood, there is evidence of persistence of the symptoms related to the pectus deformity.[15, 16, 17]

Reconstructive Surgery for Pectus Carinatum

When a pectus carinatum chest wall deformity represents a significant deviation from normal and is associated with symptoms, nonoperative or operative corrective therapy is considered reconstructive as it restores function and alleviates symptoms. When it is performed for therapeutic purposes, the surgery for pectus carinatum falls under the definition of reconstructive surgery, and not cosmetic surgery, as defined by the American Medical Association and the Centers for Medicaid and Medicare as it is "performed to improve function, but may also be done to approximate a normal appearance". [18, 19]

Overview of Therapeutic Options for Pectus Carinatum

The approach to reconstruction for pectus carinatum varies widely. Surgeons have applied operative and nonoperative techniques in treating pectus carinatum and often within each general technique of surgical approach there are variations in methodology.

1. Nonoperative therapy utilizing orthotic bracing and dynamic compression have shown equivalent outcomes when compared with operative therapy in the treatment of selected children with pectus carinatum.[20, 21, 22, 23]
2. Surgical reconstruction as an open technique involving resection of the deformed cartilages with or without sternal osteotomy has been the primary modality of therapy for several decades.[24, 25, 26]
3. Minimally invasive thoracoscopic operative reconstructive techniques with and without the resection of the deformed cartilages have recently been described, with initial excellent results.[27, 28, 29, 30, 31, 32]

Recommendations

1. For the child diagnosed with a pectus carinatum deformity physical evaluation for scoliosis should be performed.[33, 34] Dictated by the clinical presentation, an evaluation for congenital heart disease and Marfan's syndrome may also be performed.[35]
2. Symmetry of the pectus deformity, degree of sternal rotation, chest wall compliance, and the presence of a concomitant pectus excavatum deformity should be assessed. [36]
3. Although not required, chest computed tomography may assist in the surgical planning and play a role in determining the extent of the deformity in the child with a significant pectus carinatum.[20, 37, 38, 39]
4. In prepubertal children, a period of observation to follow the progression of the pectus carinatum and to allow for discussion regarding the optimal method of therapy is appropriate. Without strong evidence for ideal timing of treatment, expert opinion suggests that the age for operative therapy must be individualized, but is typically deferred until pubertal growth in nearly complete. [40]
5. As reconstructive therapy for the compliant pectus deformity, nonoperative compressive orthotic bracing is usually an appropriate first line therapy as it does not preclude the operative option. For appropriate candidates, orthotic bracing of chest wall deformities can reasonably be expected to prevent worsening of the deformity and often result in a lasting correction of the deformity. Orthotic bracing is often successful in prepubertal children whose chest wall is compliant. Expert opinion suggests that the noncompliant chest wall deformity or significant asymmetry of the pectus carinatum deformity caused by a concomitant excavatum-type deformity may not respond to orthotic bracing.[36, 41, 42, 43]
6. Open surgical reconstructive techniques are acceptable surgical options in the hands of experienced pediatric surgeons.[24, 25, 26]
7. Thoracoscopic reconstructive and other minimally invasive techniques are acceptable in some children, based on the advanced minimally invasive skills and experience of the pediatric surgeon.[27, 28, 29, 30, 21, 32]
8. Unless there is some overwhelming indication for repair, operative repair of pectus chest wall deformities is to be discouraged in children ages 5 years and younger due to the risk of disruption of normal chest wall growth with resultant chest wall restriction.[44,45, 46]
9. Expert opinion suggests ongoing evaluation through adolescence by a pediatric surgeon is appropriate in the child who has undergone nonoperative or operative chest wall reconstruction therapy. Due to rib, cartilage, and pubertal linear growth with resultant ongoing changes in the chest wall contour that may occur, the pediatric surgeon should be involved in the extended follow up of these children.

Authors

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