procedures, this is likely a result of the described intrathoracic adhesions preventing normal compensation for progressively worsening PE. Compression of the RV likely lead to the patient's exercise-induced syncope, because physical exertion increases the heart rate and decreases filling time. In the setting of diastolic impairment from compression, this results in sudden diastolic heart failure and syncope (Figure 2). After surgical correction of the PE, there was considerable relief of compression, with improvements in RV size and tricuspid valve function.

When PE occurs in the presence of CHD, surgical treatment should be considered at the time of the initial cardiac operation, even in the absence of standard indications.⁴ Such an approach has been reported with good technical results and overall outcomes. Many children with mild chest wall deformities, however, do not undergo surgical PE repair.^{2,4} Although CHD repair in early childhood does not prohibit future correction of PE, postoperative adhesions that develop at an early age may prevent the compensatory shifting of the heart that occurs during growth and can result in dramatic cardiac compression. If the PE is not

repaired at the time of the initial cardiac operation, the patient should be closely followed up with serial clinical examinations, transthoracic echocardiography, and, if needed, chest computed tomographic scans or other additional imaging modalities. Development of any cardiac compression must be identified early, because it significantly increases the risk of syncope, as seen in our patient, or even sudden cardiac death.⁵

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EDITORIAL COMMENTARY

What are the indications for pectus excavatum repair at the time of congenital cardiac surgery: Separating theory from reality

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Pectus excavatum (PE) in some form affects 0.1% of the population, with 10% to 20% of patients eventually seeking surgical consultation for repair.¹ On the basis of national practice patterns, most patients seek the consultation of a general pediatric surgeon for repair during childhood or adolescence. Thus, congenital chest wall deformities represent only a small fraction of the average thoracic surgeon's practice, leading to limited experience

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Central Message

The combination of PE and congenital cardiac defects requiring surgical repair presents a difficult therapeutic dilemma.

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with their management and operative techniques. Such limitations also extend to the full understanding of symptoms associated with PE.

Although initially faced with a dismissive attitude by the medical community, it has become clear in the latter part of the last century that even moderate cases of PE create physiologic impairment. Restrictive pulmonary defects in children are common, and cardiac problems, such as mitral valve prolapse that is reversible with PE repair, can occur in 20% of patients.² In a large 112-year retrospective autopsy series from Johns Hopkins, a correlation was evident between PE and overall survival.³ On the basis of these and other data, it is clear that repair of all but the mildest cases of PE can offer a physiologic advantage to the patient. However, when and how to repair the defect are a subject of much debate.

In this issue of the *Journal*, Dr Borrhommee and colleagues⁴ describe a case of symptomatic PE leading to recurrent syncope in a 23-year-old patient. As a result of 2 congenital cardiac operations, at 1 and 16 years of age, fixation of mediastinal structures from postoperative adhesions prevented compensatory cardiac motion away from the sternal defect in this particular patient. This resulted in severe right ventricular obstruction and exercise-induced syncope that were easily treated with an open repair after the problem was diagnosed. The authors suggest that PE repair at the time of cardiac surgery may have prevented this complication, but what are the real indications and barriers for concomitant repair and could this type of complication be avoided?

Although multiple variations of PE repair are described, almost all are based on the traditional open Ravitch approach that results in the removal of part or all of the deformed cartilage while leaving the perichondrium intact.⁵ Stabilization of the repair is required for 6 to 12 months while cartilage regrows in the proper orientation. The minimally invasive Nuss procedure achieves sternal elevation via placement and rotation of a rigid bar without concomitant cartilage resection. It thus requires internal stabilization for several years.⁶ Both procedures are well described with low morbidity in experienced hands, but the choice or necessity for an operation at the time of congenital cardiac surgery involves stout clinical decisions based on limited information.

Close to half of conservatively managed pectus defects in young children may disappear, as late as 6 years of age.⁷ No predictive algorithms exist that allow for determination of future pectus severity. Thus, on the basis of chance alone, many toddlers may not require repair even if a defect occurs

at the time of cardiac operation. The Nuss technique, although having many advantages over the open approach, may present an impediment to proper sternal alignment and healing immediately after median sternotomy, although the combination of these 2 procedures has been described.⁸ The placement of a rigid bar may interfere with productive cardiopulmonary resuscitation,⁹ and the necessity of Nuss bar maintenance for several years may interfere with any future plan for a staged repair. The Ravitch technique, which requires costal cartilage resection, may lead to excessive blood loss due to postcardiotomy coagulopathy and may hinder sternal healing due to disruption of sternal perforators or thoracic vessels. Because of these concerns, some have recommended a staged approach for PE and congenital heart repair.¹⁰ Furthermore, excessive cartilage resection or disruption of growth plates during an open Ravitch procedure in young patients may result in restrictive asphyxiating thoracic dystrophy or acquired Jeune syndrome.¹¹ In view of these data, the decision to wait until adolescence, or completion of growth, before repairing PE may be prudent on multiple fronts. Thus, in theory, the decision of Borrhommee and colleagues⁴ to avoid PE repair at the time of infundibular stenosis resulted in syncope and reoperation in this particular patient. In reality, their avoidance of concomitant repair may have been based on solid clinical judgment.

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