Journal of Pediatric Surgery (2012) 47, E61–E64

ELSEVIER

Journal of Pediatric Surgery

www.elsevier.com/locate/jpedsurg

Development of supravalvular pulmonary artery stenosis following a Nuss procedure

Lynnette Mazur*, Lisa de Ybarrondo, Laurens Pickard, P. Syamasundar Rao

Received 18 June 2012; revised 26 September 2012; accepted 28 September 2012

Key words:
Pectus excavatum;
Nuss repair;
Pulmonary artery stenosis

Abstract We report a case of a 13-year old girl with pectus excavatum who had a Nuss procedure and two years later a new cardiac murmur appeared which on investigation was diagnosed as supravalvular pulmonary artery stenosis. Following removal of the Nuss bar the stenosis resolved.

Pectus excavatum (PEx) is a deformity of the anterior chest wall with posterior indentation of the sternum and adjacent costal cartilages. The prevalence is approximately one in 300 to 400 live births with a male to female preponderance of 4:1. While the indications for surgery are not clearly defined, recent reviews suggest presence of two or more criteria listed in their tables [1,2]. A variety of surgical techniques have been used to correct the deformity; the two most commonly employed techniques are those described by Ravitch [3] and Nuss [4]. In the Nuss procedure, a steel convex bar is inserted and eventually positioned such that the convexity faces anteriorly, thereby addressing the chest deformity. The bar is removed two years following placement by which time permanent chest remolding is deemed to occur. The objectives of this report are to illustrate a rare cardiac complication associated with the Nuss bar and to review long term complications of this approach to treat PEx.

1. Case presentation

MC, a 13 year old white female with pectus excavatum had an open repair at age 3 years and a Nuss procedure at 11 years. Prior to planned removal of the bar two years later, a cardiac murmur was noted during pre-anesthesia assessment leading to referral for cardiac evaluation. Her only complaints were a sharp, pinching, left-sided chest pain that occurred only with sneezing for the past several months and an inability to sleep supine since the time of the bar placement. She denied any history of chest trauma. On physical examination her height, weight, and body mass index (BMI) were 159 cm (50th–75th percentile), 45 kg (25th–50th percentile), and 18 (25th percentile) respectively, similar to the percentiles at the time of placement of the bar. Her cardiac exam revealed normal cardiac impulses, no thrills, normal first heart sound at the apex, single second heart sound at the left upper sternal border, a long, grade 3/6 ejection systolic murmur at the left mid-sternal border that peaked late in systole and radiated to the infraclavicular regions and axillae, no diastolic murmurs or signs of heart failure. The remainder of the physical exam was unremarkable.

The chest x-ray (Fig. 1A) showed the cardiac size to be at the upper limits of normal with a slight prominence of the right ventricular outflow tract (arrow in Fig. 1A) which is slightly more prominent than in the x-ray performed on the day of surgery (Fig. 1B). It also showed that the Nuss bar had shifted (Fig. 1A & B). An electrocardiogram (ECG) revealed a normal sinus rhythm with right axis deviation and an RSR prime pattern in the right chest suggesting right ventricular hypertrophy. Echocardiogram revealed normal

* Corresponding author. Shriners Hospital for Children, Houston, Texas 77030, USA. Tel.: +1 713 793 3749; fax: +1 713 793 3779.
E-mail address: Lynnette.j.mazur@uth.tmc.edu (L. Mazur).

0022-3468/$ – see front matter © 2012 Elsevier Inc. All rights reserved.
http://dx.doi.org/10.1016/j.jpedsurg.2012.09.049

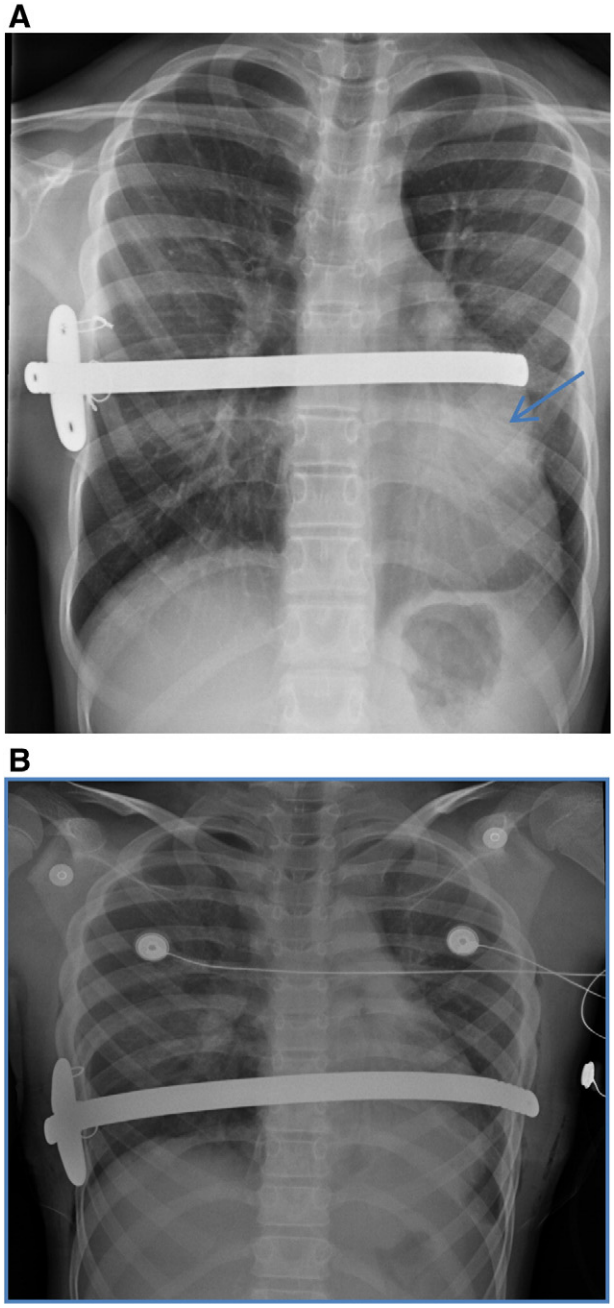

Fig. 1 Chest roentgenograms in posterior–anterior view demonstrating prominent right ventricular outflow tract (arrow in A) which was absent in a chest x-ray obtained on the day of bar placement (2B).

sized left ventricle with normal function and moderate enlargement of the right ventricular cavity and increased thickness of the right ventricular wall. There appeared to be echo-dense structures within and outside the pulmonary artery (Fig. 2A), causing severe obstruction with an estimated peak instantaneous Doppler gradient of 94 mmHg and a mean of 56 mmHg (Fig. 2B &C), suggesting that the obstruction was severe. The supravalvular pulmonary artery narrowing (stenosis) was thought to be due to compression of the pulmonary artery by the Nuss bar. Discussions by the surgeons and the cardiologist concluded with the recommendation of removal of the bar at a tertiary care children's hospital where cardiopulmonary bypass was available. The bar was removed by positioning the patient in the lateral thoracotomy position and by opening the old right side incision site, releasing the anchor plate, cutting and removing all wires and gently pulling the bar out along the curvature of the implant. There were no complications.

Five months after removal of the Nuss bar, pediatric cardiology evaluation revealed improved symptomatology regarding the chest discomfort and a grade 2/6 early diastolic decrescendo murmur best heard at the left upper and left midsternal borders; the systolic murmur had resolved. An ECG revealed increased S-waves in the left chest leads, consistent with mild right ventricular hypertrophy. An echocardiogram revealed a mildly dilated right ventricle and normal left ventricular size and function. There was minimal residual gradient across the right ventricular outflow tract (peak instantaneous gradient of 15 mmHg) and mild to moderate pulmonary insufficiency (Fig. 3). The disappearance of the systolic murmur was thought to be related to elimination of the pulmonary artery obstruction and the appearance of the diastolic murmur was related to distortion of the pulmonary valve leaflets by the Nuss bar and supravalvular pulmonary artery stenosis close to the pulmonary valve.

2. Discussion

The Nuss procedure, introduced in 1997, has the advantages of being minimally invasive and producing excellent cosmetic results. However, bar displacement can occur and is one of the most common complications with a prevalence up to 16.6% [5–8]. Other late complications include subcutaneous emphysema [9], pericardial effusion [9], erosion of the bar into the internal mammary artery [10], persistent thoracic outlet syndrome [11], overcorrection or progression to pectus carinatum [7], cardiac tamponade [12] and ossification around the Nuss bar [9]. However, the authors were unable to find any reports on pulmonary artery stenosis related to the Nuss procedure.

The patient in this paper developed supravalvular pulmonary artery stenosis, detected two years after surgery. While the exact cause of this complication is not known, it may be postulated that displacement of the Nuss bar along with natural growth of the heart resulted in compression of the pulmonary artery with consequent development of severe supravalvular pulmonary artery stenosis. The stenosis is well documented in the echo-Doppler studies (Fig. 2) which almost completely resolved following the removal of the bar (Fig. 3). The development of pulmonary insufficiency may be related to distortion of the pulmonary valve leaflets, causing their inability to coapt well to have a competent valve.

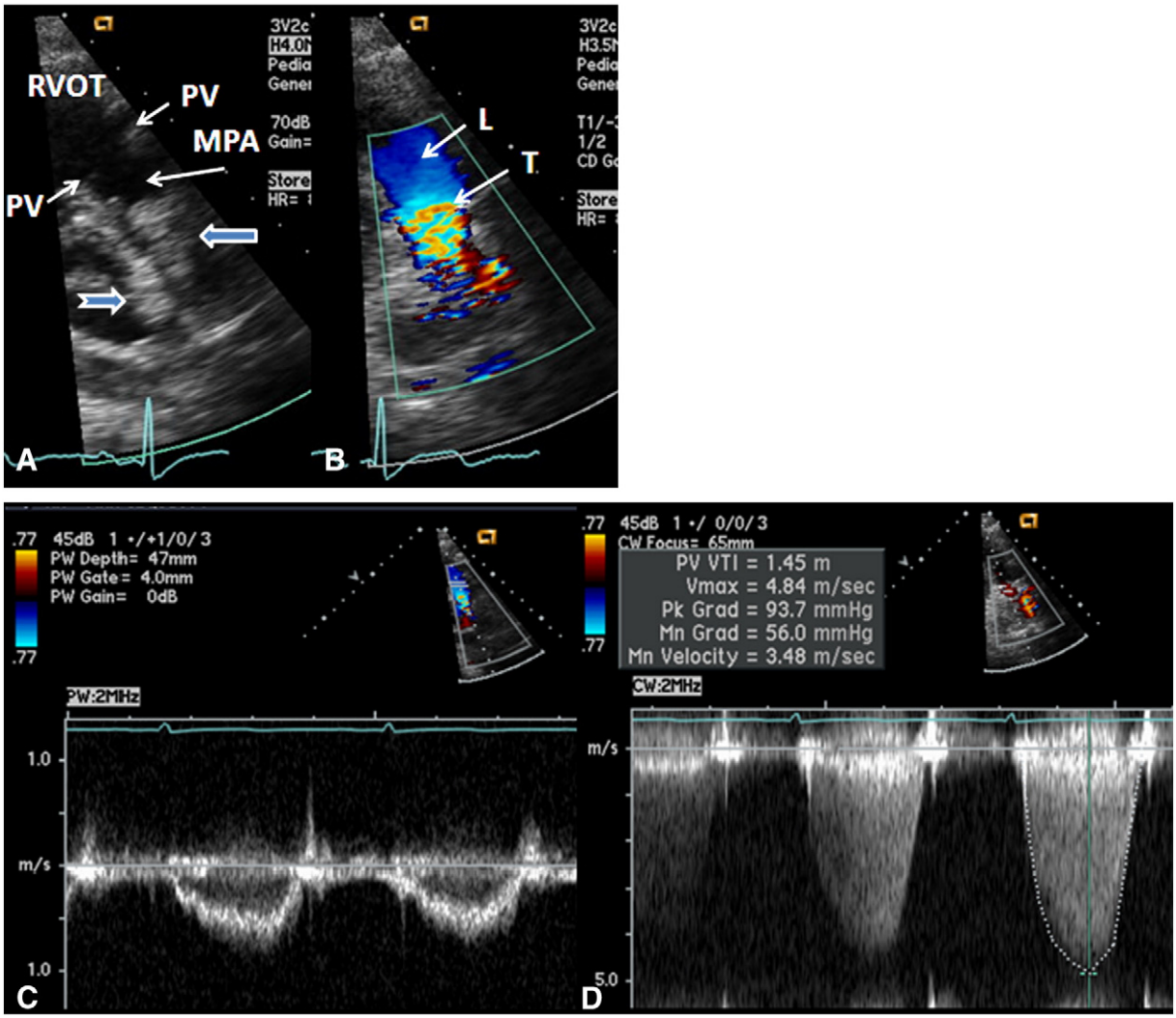

Fig. 2 (A) Selected video frame from a precordial short axis view of the heart demonstrating echo dense structures (thick blue arrows) within and outside the main pulmonary artery (MPA). Pulmonary valve (PV) leaflets (small arrows) are shown and appear normal. The right ventricular outflow tract (RVOT) and proximal MPA are free of any echo-dense structures. (B) Color Doppler mapping of the same structures as in panel A shows normal laminar (L) flow in the RVOT and proximal MPA and turbulent (T) flow starting in the MPA, indicating obstruction. (C) Pulse Doppler sampling from the proximal MPA which shows normal flow velocity. (D) Continuous wave Doppler sampling demonstrating high velocity flow across the MPA with a calculated peak instantaneous gradient of 93.7 mmHg and a mean gradient of 56 mmHg, indicating severe obstruction.

Recommendations for bar placement vary. One study recommended "Bar stabilization and fixation … on the left" [7] while another stated that the "bar should be secured with two stabilizers…" [8]. In reviewing our references that included radiographs, stabilizers were noted on the right in three [5,10], on the left in one [9], and bilateral in one [8].

Based on our observation of development of supravalvular pulmonary artery stenosis following the Nuss procedure, we would recommend careful auscultation by the primary care physician during routine well-child care visits and if a significant or new cardiac murmur is detected, cardiac evaluation including echo-Doppler studies may be warranted. Further studies on the risks and benefits and on which side to place the stabilizer are needed.

References

[1] Colombani P. Preoperative assessment of chest wall deformities. Semin Thorac Cardiovasc Surg 2009;21:58-63.
[2] Jaroszewski D, Notrica D, McMahon L, et al. Current management of pectus excavatum: a review and update of therapy and treatment recommendations. J Am Board Fam Med 2010;23(2):230-9.
[3] Ravitch M. Pectus excavatum and heart failure. Surgery 1951;30:178-82.
[4] Nuss D, Kelly RE, Croitoru DP, et al. A 10-year review of a minimally invasive technique for the correction of pectus excavatum. J Pediatr Surg 1998;33:545-52.
[5] Hebra A, Swoveland B, Egbert M, et al. Outcome analysis of minimally invasive repair of pectus excavatum: review of 251 cases. J Pediatr Surg 2000;35:252-8.
[6] Castellani C, Schalamon J, Saxena AK, et al. Early complications of the Nuss procedure for pectus excavatum: a prospective study. Pediatr Surg Int 2008 Jun;24(6):659-66.

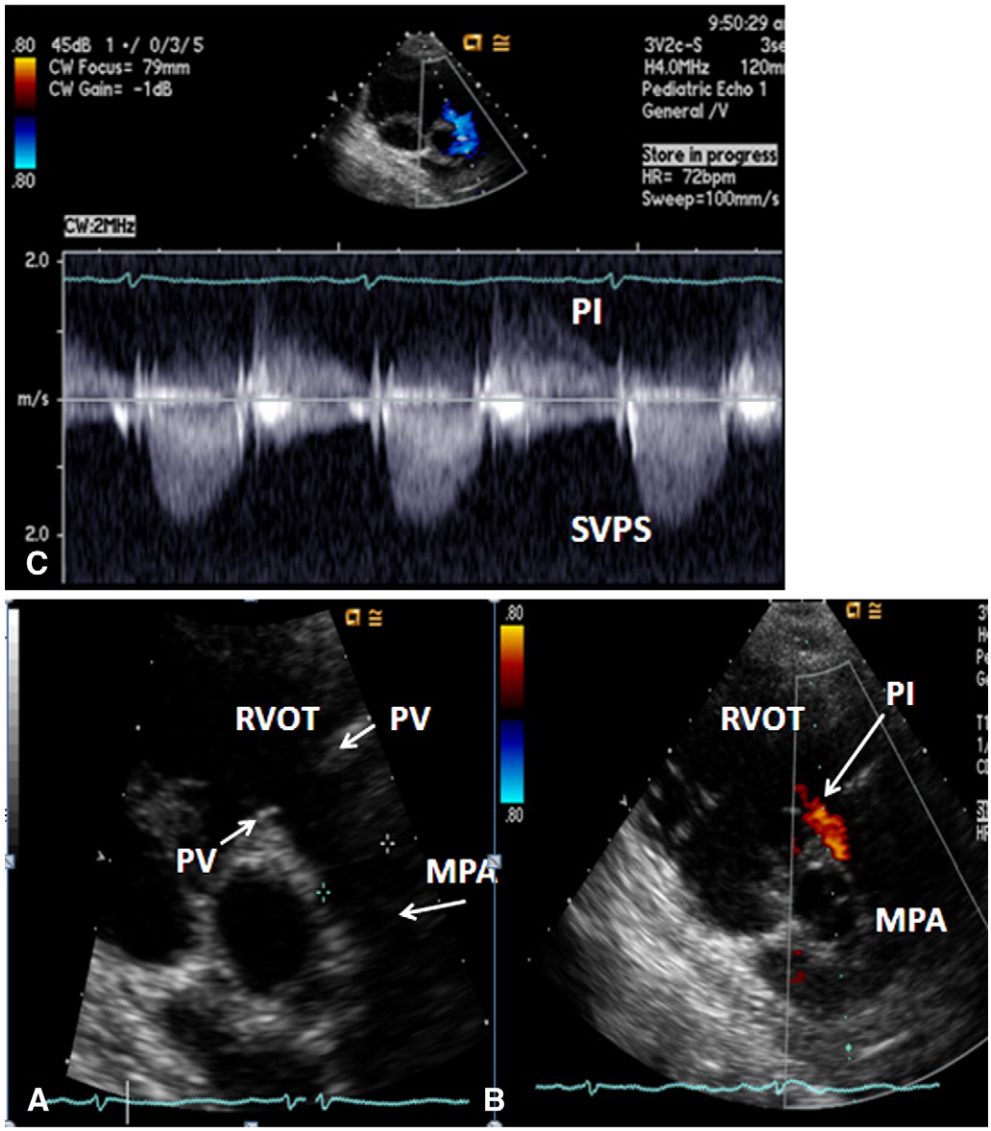

Fig. 3 Echo-Doppler studies performed five months after removal of the Nuss bar. (A) Selected video frame from a precordial short axis view of the heart demonstrating no echo dense structures in the right ventricular outflow tract (RVOT) and main pulmonary artery (MPA). Pulmonary valve (PV) leaflets (arrows) are shown. (B) Color Doppler mapping of the same structures as in panel A shows pulmonary insufficiency (PI) (arrow). (C) Continuous wave Doppler sampling demonstrating low Doppler flow velocity across the MPA with a calculated peak instantaneous gradient of 15 mmHg, indicating minimal supravalvular pulmonary stenosis (SVPS) and pulmonary insufficiency (PI).

[7] Kelly RE, Goretsky MJ, Obermeyer R, et al. Twenty-one years of experience with minimally invasive repair of pectus excavatum by the Nuss procedure in 1215 patients. Ann Surg 2010;252:1072-81.
[8] Tedde ML, Mianez de Campos JR, Das-Neves-Pereira JC, et al. The search for stability: bar displacement in three series of pectus excavatum patients treated with the Nuss technique. Clinic 2011;66:1743-6.
[9] Vegunta RK, Pacheco PE, Wallace LJ, et al. Complications associated with the Nuss procedure: continued evolution of the learning curve. Am J Surg 2008 Mar;195(3):313-6.
[10] Adam LA, Lawrence JL, Meehan JJ. Erosion of the Nuss bar into the internal mammary artery after 4 months after minimal invasive repair of pectus excavatum. J Pediatr Surg 2008;43:394-7.
[11] Lee SH, Ryu SM, Cho SJ. Thoracic outlet syndrome after the Nuss procedure for the correction of extreme pectus excavatum. Ann Thorac Surg 2011;91:1975-7.
[12] Yang MH, Cheng YL, Tsai CS, et al. Delayed cardiac tamponade after the Nuss procedure for pectus excavatum: a case report and simple management. The Heart Surgery Forum #2007-1217.