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Dr. Wenlin Wang
Professor, Department of Chest
Wall Surgery, Guangdong
Second Provincial General
Hospital, Guangzhou, China

Dr. Weiguang Long
Associate Professor, Department
of Chest Wall Surgery,
Guangdong Second Provincial
General Hospital, Guangzhou,
China

Dr. Yang Liu
Resident Doctor, Department of
Chest Wall Surgery, Guangdong
Second Provincial General
Hospital, Guangzhou, China

Dr. Bin CAI
Resident Doctor, Department of
Chest Wall Surgery, Guangdong
Second Provincial General
Hospital, Guangzhou, China

Dr. Juan Luo
Resident Doctor, Department of
Chest Wall Surgery, Guangdong
Second Provincial General
Hospital, Guangzhou, China

Corresponding Author:
Dr. Wenlin Wang
Professor, Department of Chest
Wall Surgery, Guangdong
Second Provincial General
Hospital, Guangzhou, China

Reoperation after failure of Nuss procedure on Wenlin chest with Poland syndrome

Dr. Wenlin Wang, Dr. Weiguang Long, Dr. Yang Liu, Dr. Bin CAI and Dr. Juan Luo

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Abstract

Wenlin chest is a rare congenital thoracic deformity, which may be combined with other deformities. Because its main lesions are the protrusion and depression of the anterior chest wall, if only one kind of lesions is treated, the deformity will not be corrected completely, and the operation may fail. We met a 20-year-old female patient with Wenlin chest and Poland syndrome who underwent Nuss procedure two years before. The operation was unsuccessful, and the patient still had serious deformity. We performed the reoperation for her, and satisfactory results were obtained.

Keywords: Wenlin chest, Poland syndrome, Matrix RIB, pigeon breast, pouter pigeon chest, pectus arcuatum, currarino-Silverman syndrome, chondromal deformation, reoperation, Nuss procedure

Introduction

Wenlin chest is a rare thoracic deformity named by us [1, 2]. Because the anterior chest wall protrusion is the main pathological feature of it, it was regarded as a kind of pectus carinatum in early years [3]. With the deepening of the understanding of this deformity, more characteristics are recognized, thus other names were gradually appearing in clinic. These names included: pouter pigeon chest, pigeon breast, pectus arcuatum, Currarino-Silverman syndrome and chondromal deformation, etc [4-6]. Our department is an independent chest wall surgery department. One of our main works is to complete various thoracic deformity operations. In our work, we have treated many patients with this deformity [7-9]. In order to facilitate our own work, we also named this deformity that is Wenlin chest [1, 2]. Since this deformity is usually a congenital disease, it may be combined with other deformities. We had met a special Wenlin chest patient with typical Poland syndrome. Before coming to our hospital, she had a failed Nuss procedure in local hospital two year before. We performed the second operation for her and got satisfactory results.

Case Report

The patient was a 20-year-old female. Before the age of 10, the patient's chest wall had no obvious abnormality, but then gradually developed a median protrusion of the anterior chest wall, which was not obvious on the right side of the chest wall. Meanwhile, she also had slight depression below the median part of the protrusion. After puberty, the deformity worsened. Her right chest wall gradually depressed, and there was obvious dysplasia on the right breast, but the left chest wall and left breast was basically normal. The patient visited local hospital when he was 18-year-old. Diagnosed as pectus excavatum, Nuss procedure was performed for her, but the operation was not successful. The deformity still existed without any change after operation. In order to treat the deformity completely, the patient was admitted to our hospital at the age of 20 for reoperation. Preoperative physical examination found that the anterior chest wall was centrally protrusive, with depression just below the protrusion; the chest wall was asymmetrical, with normal development of the left breast but dysplasia of the right breast; and surgical scars could be found on both sides of chest wall (Fig. 1). Preoperative imaging examination showed that the upper part of the anterior chest wall was protrusive, while the lower part is depressed in the middle; the sternum is obviously thickened; the right 3rd, 4th

and 5th costal cartilages was absent; and there were two steel bars and fixation plates in the chest (Fig. 2, 3). The patient was diagnosed as Wenlin chest with Poland syndrome before operation. After full preoperative preparation, the operation was performed under general anesthesia. The main contents of the operation consisted two parts: the first is to take out the steel bars and plates, and the second is to implement the re-correction of the chest wall. Two incisions at the scars on the lateral chest wall were performed respectively to expose both ends of the steel bars and plates. After the fixation wires were removed, the steel bars and plates were taken out. An incision was made in the middle of the anterior chest wall to expose the protruding sternal angle and sternal body. A bone rongeur was used to bite off the most obvious part of the protruding sternum and retain the posterior cortex of the sternum. The bony structures of the anterior chest wall were exposed, and the deformity was corrected with two MatrixRIBs (Fig. 4). The protrusive part of the sternum was compressed by the middle part of the two MatrixRIBs, with other parts of them were fixed on the ribs. The defect of the right chest wall was repaired by the right half of the MatrixRIBs. After the MatrixRIBs were firmly fixed, the incisions were closed and the operation was completed (Fig. 5). There was no complication during the operation. X-ray examination after the operation showed that the position of MatrixRIBs was normal and the shape of the bone structures of the chest wall was satisfactory (Fig. 6). The patient was discharged 9 days after operation. Follow up for 1 year after operation, CT examination showed that the positions of MatrixRIBs were satisfactory, and the chest wall deformity did not reoccur (Fig. 7).

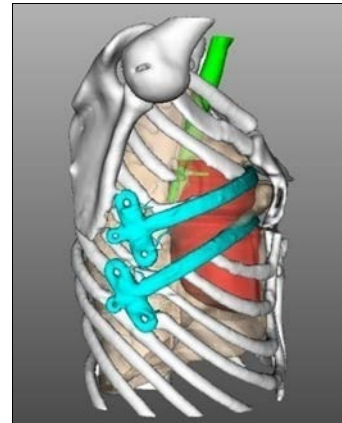


Fig 3: Three dimensional picture of chest wall. The anterior chest wall is protrusive and depressed, and the sternum is thickened in an "S" shape. The positions of the steel bars are abnormal



Fig 4: Surgical picture. The locations of the two MatrixRIBs are displayed

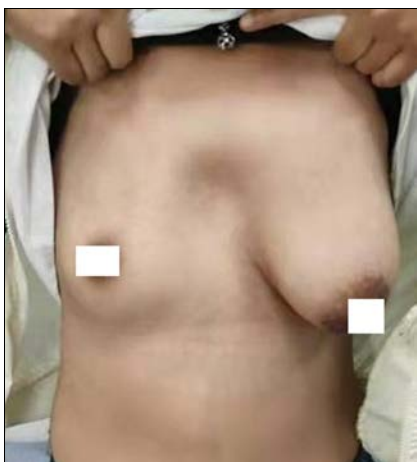


Fig 1: Appearance of chest wall before operation



Fig 5: Appearance of chest wall after operation

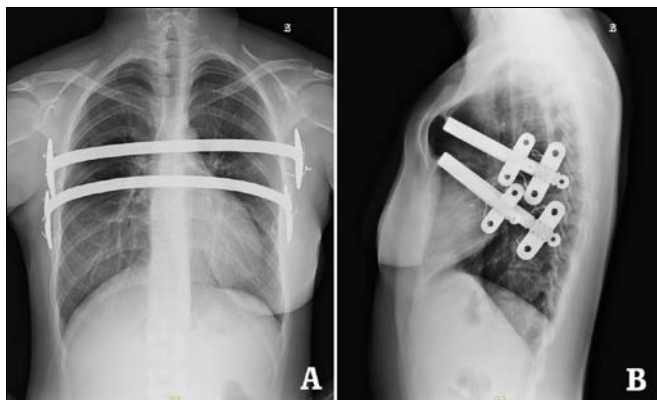


Fig 2: Chest X-ray examination before operation. A. Front view. Two steel bars and fixed plates can be seen, and the positions seem normal; B. Side view. The steel bar positions have obvious problems



Fig 6: Postoperative X-ray examination. The locations of the two MatrixRIBs are displayed

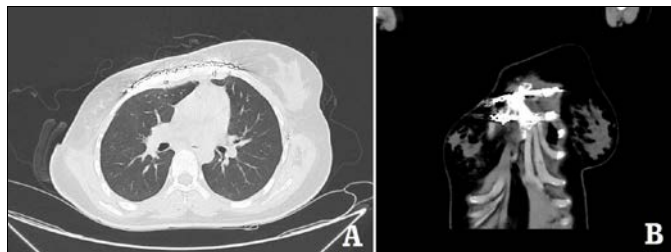


Fig 7: CT examination one year after operation. A. On the horizontal plane, the bone structure contour of the thorax is basically normal; B. On the coronal plane, the MatrixRIBs are in the same plane of the sternum.

Discussion

Wenlin chest is a rare thoracic deformity [3-6]. So far, the total number of cases reported worldwide does not exceed 100. One of the most important features of this deformity is the prominent protrusion of the anterior chest wall [1, 2]. Because of this, the deformity has always been regarded as a kind of pectus carinatum [3]. However, with the progress of cognition, other features of this deformity have been gradually taken seriously, thus various names have emerged, and this deformity is no longer just regarded as pectus carinatum [3-6].

Essentially, Wenlin chest is a complex deformity, with both protrusion and depression. The protrusion is located near the sternal angle and the depression is located directly below it. This kind of deformity is completely different from the general compound deformity, mainly due to the structure of the sternum [1, 2]. Since the sternum of Wenlin chest is obviously thickened and extremely hard, sandwich surgery for common complex deformities cannot be used for the correction of Wenlin chest [10]. It can be seen that in order to complete the treatment of Wenlin chest, the sternum must be treated first. The previous popular method is generally open surgery, namely Ravitch procedure [3-6]. In this procedure, one of the target structures is the sternum. In the operation we designed, we also aim at the sternum first, but this is not the whole operation. We call it pre-shaping, which is the basis of the total operation [11]. With such a basis, some minimally invasive surgical procedures can be easily implemented. We usually use a combination of Wenlin procedure [12-16] and Wung procedure [17-18] or Wang procedure [19-23] in the treatment of this deformity, which is not a sandwich surgery, but can achieve perfect results.

Poland syndrome is a common thoracic deformity [24]. Its lesions involve two sorts of structures at the same time, namely, the bone structures and soft tissues of the chest wall. The pathological changes of bone structures can be manifested as dysplasia or depression of unilateral ribs or costal cartilages, and severe patients may have the defect of these structures. Three costal cartilages were absent on the right side of this patient, which made the lesion a serious type. The soft tissue lesions are mainly the developmental disorders of chest muscles and breast. In this case, the pectoralis major muscle is poorly developed, and the breast is basically not developed. From the appearance, the chest wall of the patient is obviously asymmetrical. This has become an important reason for her to undergo surgery. Theoretically, the treatment of Poland syndrome requires simultaneous correction of bone structures and soft tissues. However, due to their different development phases, different structures can be corrected at different times.

Poland syndrome is a congenital chest wall deformity, which may exist with other deformities. This patient has both Poland syndrome and Wenlin chest. This is an extremely rare

combination and has not been reported before. The coexistence of the two lesions make the deformity complex, so the possibility of being misdiagnosed increases. This patient was once diagnosed as pectus excavatum, which is obviously a very regrettable diagnosis. When Nuss procedure was performed on the depression of this patient [25], because the sternum was too hard, it was almost impossible for the steel bar to change the shape of the depression, so the operation was bound to fail.

This patient is a combined deformity of Wenlin chest and Poland syndrome. In addition to the obvious lesions of the sternum, there is a defect of costal cartilages on the right chest wall, so it needs to be corrected at the same time. The operation should consist two parts: one part is deformity correction, and the other is defect reconstruction. Because the reconstruction materials may not be taken out after the operation, we chose MatrixRIB for surgery. This material can be permanently retained in the human body, so it is suitable for reconstruction of defects [26]. Since the material has enough mechanical strength, we also used it in the correction of median protrusion, and achieved satisfactory results.

Conclusion

Wenlin chest is a congenital thoracic deformity, which may be combined with other thoracic deformities. These deformities should be corrected at the same time. For Wenlin chest, because the sternal lesion is the root of the deformity, it must be managed at first. Without the pre-shaping on sternum, the operation is doomed to failure.

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Author's Contribution

Not available

Conflict of Interest

Not available

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