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OUR EXPERIENCE IN THE MANAGEMENT OF CONGENITAL CHEST WALL DEFORMITIES

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SUMMARY – Chest wall deformities are relatively rare diseases of unknown etiology, which occur in childhood and adolescence. Pectus deformities show familial occurrence with very rare spontaneous resolution. Operative treatment is one of the possible therapeutic options for deformity correction by classic operative procedure or by minimally invasive method of treatment (endoscopic). There is no consensus among surgeons about the age at which correction of the chest wall deformity should best be performed because therapeutic results are very good irrespective of the method of treatment employed. Therapeutic results in 105 patients operated on by the classic method during the 1985-2005 period at University Department of Thoracic Surgery, Jordanovac University Hospital for Lung Diseases in Zagreb, Croatia, are reported.

Key words: *Funnel chest – pathology; Funnel chest – surgery; Funnel chest – therapy; Surgical procedures operative – methods*

Introduction

Pectus excavatum or funnel breast is the most common form, and pectus carinatum or chicken breast by far less common form of chest wall deformity. Both defects are congenital anomalies of the anterior thoracic wall, however, the true reason for their occurrence has not yet been fully clarified. It has been postulated that these deformities occur due to defective growth of the cartilaginous segments of the ribs, which push the sternum ventrally or dorsally. Initial changes occur in the early childhood to become more prominent with the child's growth. Funnel breast is characterized by a shallow or deeper depression in the lower half or lower two thirds of the sternum, with the deepest point above the xiphoid process. It is mostly accompanied by a more or less pronounced unilateral or bilateral deformity of the costal cartilages, from the third to seventh or eighth rib. Costal arch deformity may also be occasionally present. Considering all these deformity combinations, some authors classify pectus excavatum into ten grades, from the mildest form of purely cosmetic defect (grade I) through pronounced kyphoscoliosis with bilateral costal arch deformities and deep depression in the sternum area (grade X). In half of these patients, the sternum is asymmetric with longitudinal or transverse curvature. Major deformities may frequently be associated with pressure upon the medially positioned organs of the mediastinum, primarily the heart and great blood vessels, and to a certain extent upon the lungs. This in turn may result in cardiac abnormalities such as arrhythmia, tachycardia, cardiac murmur, 10%-30% restriction in lung capacity, limited physical activity and fatigability, effort intolerance, etc. Disturbances can be verified by electrocardiography (ECG), spirometry, sternum x-ray in anteroposterior and lateral projection, and by computed tomography (CT) of the thorax as needed.

Pectus excavatum is found in one *per* 500-1000 children, usually in families with a positive history, and is frequently associated with other musculoskeletal defects

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such as scoliosis, which is present in one fifth of these patients. Alteration is generally observed short after birth or in early childhood, to progress slowly until the end of the child's growth. Regression is extremely rarely observed because in adolescence the cartilages, bones and ligaments are fixed in the pathologic position.

Initially, the symptomatology may not be strongly pronounced until the children get exposed to major physical strain at day-care centers and especially at school. Then, psychic burden upon the child with such a deformity is first noticed, manifesting by the child's isolation from the company of his/her school-mates, introversion, tending to conceal the physical defect from the others, avoiding swimming, taking shower and other activities in company with others, etc.

In case of major deformities, respiratory difficulties develop in the form of tachypnea and excessive excursions of the respiratory diaphragm to compensate for the inactivity of the pathologically altered part of the sternum. All this leads to fatigability and effort intolerance as compared with healthy peers. An increased incidence of respiratory infections and asthma may also be occasionally recorded.

In comparison with pectus excavatum, the incidence of pectus carinatum is considerably lower, affecting one *per* 3000-5000 children; however, like the former, also showing a 3:1 male to female ratio. There are three types of sternum protrusion: type I (keeled chest); type II (pigeon chest); and type III (asymmetric or lateral pectus carinatum)[1].

There are continuing controversies among the professionals about the age at which the planned deformity correction should best be done. Some believe that optimal timing is by age 4, others advocate age 8, while some consider it is best be performed upon the growth completion. Yet, what is to be done with the serious psychical problems in these young individuals? Is there any warranty that the defect will not recur postoperatively irrespective of the age at which the correction is performed? What method of treatment to choose?

There are a number of therapeutic options for sternum deformities: plastic surgery with presternal implants for defect correction; minimally invasive surgery for implantation of retrosternal metal arches; corrective sternum osteotomy; and subchondral cartilage ablation with or without temporary retrosternal supportive plates according to Ravitch and many other authors[2].

The management of chicken breast does not require placement of supportive plates, although the operative procedure greatly resembles funnel chest correction. The procedure takes from one to five hours, depending on the type of operation, with 4- to 7-day hospital stay. Currently, early mobilization, on postoperative day 2, and light physical therapy including breathing exercises and contraction of the upper and lower extremity musculature on postoperative day 1 are preferred. Exemption from physical efforts is recommended for 4-6 months, and plate removal at 8-12 months postoperatively[3].

Patients and Methods

During the 1985-2005 period, 105 patients (92 male and 13 female) were operated on for chest wall deformities at University Department of Thoracic Surgery, Jordanovac University Hospital for Lung Diseases. Preoperative work-up included thorax x-ray and lateral sternum projection, ECG and laboratory tests. In 14 male patients with severe chest wall deformities, pronounced cardiorespiratory impairments and physical effort intolerance, spirometry, thorax CT, heart ultrasonography (US) and cardiologist examination were also performed. Other patients suffered psychical problems related to their cosmetic defects, with some elements of social phobia.

The operative method of treatment according to Ravitch, in recent years modified according to Welch, has been used for years at our department. Compared with the original method, where patient was bed-ridden for three weeks without mobilization, the modified method allows for patient mobilization on postoperative day 3, and discharge from the hospital on postoperative day 7-8 if free from complications. This method requires the use of a temporary retrosternal plate to ensure chest stability from the very beginning, along with early rehabilitation and pain reduction in the operative area.

The procedure is initiated by vertical incision above the sternum or by transverse arcuate incision below the two mammillae with craniad convexity. This is followed by subcutaneous tissue separation from the pectoral muscle and rectus abdominis muscle, then these muscles are also prepared and separated from the thoracic wall and costal arches. Now costal cartilages are seen and peeled off subchondrally to leave perichondrium alone; the procedure is extended to costal arches if involved by the defect. Then, intercostal muscles and perichondrium are separated from the sternum by the cauter, blunt preparation is employed to reach below the xiphoid and separate pericardium from the sternum. In the anterior surface of the sternum, a cuneiform cut is

made by a chisel at the site where the corpus is sagging into the depression, and the posterior cortex is broken anteriorly to achieve due correction. The procedure should be performed with great caution, to prevent full detachment or breaking of the sternum, which would then heal with difficulty unless there is at least one intact corticalis left. Then perichondrium remnants are sutured to form canals at the site of the cartilage removed, and then are resutured to the sternum edge. Supportive plate is previously shaped and now placed under the sternum to keep the entire construction in the desired position. Redon drain is placed behind the sternum, while rectus muscle and pectoral muscle are sutured in the Mercedes star fashion. The subcutaneous tissue and the skin are sutured by the intracutaneous technique. Drain is removed on the next day, and unresorbed skin sutures are removed on day 10.

Results

From 1985 till 2005, 105 patients were operated on at our department for chest wall deformities using classic method according to Ravitch or its modification according to Welch. There were 92 (87.6%) male and 13 (12.4%) female patients, with a rate ratio of 7.08; 95% CI 4.06-13-16; $p<0.001$. The patients were aged 5-37, mean (standard deviation, SD) 17.9 (5.6) years, with no statistically significant sex difference: male 5-35, mean (SD) 17.7 (0.6) and female 5-37, mean (SD) 19.5 (1.5) years; $F=1.26$; $p=0.264$. According to type of chest wall deformity, there were 76 (72.4%) cases of pectus excavatum and 29 (27.6%) cases of pectus carinatum, with a rate ratio of 2.62; 95% CI 1.72-4.07; $p<0.001$. There was no statistically significant sex difference in the rate of either chest wall deformity. In male patients, pectus excavatum was present in 69 (75%) and pectus carinatum in 23 (25%) cases. In female patients, the respective figures were 7 (53.8%) and 6 (46.2%); $\chi^2=2.55$; $p=0.1104$; risk ratio 1.39; 95% CI 0.32-6.15, yielding no significant difference from the rate reported by Welch[6] ($p>0.99$; Fisher exact test). The complications included wound infection and hemorrhage in 4 (3.8%) patients each, and pneumothorax in 6 (5.7%) patients. In total, 14 complications were recorded in the patient series (13.3%; 95% CI 7.8-20.9), i.e. threefold that reported by Welch[6] (risk ratio 3.03; 95% CI 1.67-5.5; $p=0.002$; Fisher exact test). The mean length of hospital stay was 12 days while operating on according to the original Ravitch method, and was reduced to 7 days when switching to the modified method according to Welch.

Discussion

As sparing methods of treatment have recently been preferred, the method according to Nuss has been increasingly used in the last decade, and has been well accepted by pediatric as well as adult patients. The main feature of the method first presented in 1998 is that it requires no resection of the cartilaginous segments of the ribs or any procedure on the sternum itself. Using an elastic metal plate placed by use of a video thoracoscope, the anterior thoracic wall is being pushed anteriorly over 2-3 years, when a satisfactory correction of the deformity is achieved[4].

Long-term results of the method are yet to be properly validated, in comparison with the method of Ravitch and numerous modifications of the original method in particular. Some studies are already under way and their number will certainly increase with time. These authors report on the operating time to be significantly reduced from the original method to 2 hours and postoperative hospital stay to 4 days, with a minimal rate of complications and excellent results recorded earlier in the postoperative period in comparison with the method of Nuss. Patients showed high satisfaction with the results also at long-term follow up, while reoperation was required in a negligible proportion of patients operated on[5].

In a series of 704 patients, Welch and Shamberger recorded 107 cases of associated scoliosis, 4 cases of kyphosis, and several cases of rare syndromes like Marfan or Pierre Robin syndrome. Positive family history of thoracic wall deformity was present in 37% of patients, 11% of these with a positive history of familial scoliosis. These authors report on a low rate of complications (4.4%) and even less relapses (2.7%), whereas reoperation was required in only 12 patients[6].

There are many other modifications of the original Ravitch method, which implied a several-hour operating procedure. So, Fonkalsrud and Mendoza reduced the procedure from 4-5 hours to about 2 hours employing a similar technique of cutting the cartilaginous processes of the ribs at two sites (at costochondral junction and at costal cartilage attachment to the sternum). Patient follow up demonstrated the method to be as useful as the original one, yielding very good long-term results. Only five of 275 patients operated on were not satisfied with the operative result, and retrosternal plate was removed at 6 months. The mean length of hospital stay was 2.9 days, postoperative pains were mild to moderate, and

all patients resumed their daily activities soon after the procedure[7].

Results obtained by the less aggressive method of treatment with endoscopic placement of retrosternal plate should definitely be paid due attention, not only for operative time reduction to less than one hour on an average (25-130 minutes) but also for the low rate of intraoperative and postoperative complications (4.1% and 9.3%, respectively). The mean length of hospital stay was 5.3 days, while plate removal was performed at a mean of 26 (range 22-32) months.

Additional resection of protruding cartilages performed in the same act with supportive plate removal was required in only 3.5% of patients. Excellent results were achieved in patients below age 12, whereas a modification of the original method such as placement of two plates or possible rib resection and sternum osteotomy was occasionally required in older patients. In 99% of patients, the operation was indicated for cosmetic defect, while pronounced effort intolerance and fatigability were present in half of the patients. Cardiac axis deviation and impaired repolarization were detected on ECG in 62.9% of patients, while heart US revealed mitral valve prolapse in as many as 59% of patients. Pulmonary function tests showed restrictive ventilation impairments in 38.6% of patients[8].

Along with these congenital chest wall deformities, acquired deformities consequential to various pathologic events within and on the thoracic wall have been increasingly reported. Fokkin and Robicsek classify these deformities into four groups as follows: group 1, pathologic process within the chest (mediastinal tumors, cardiac hypertrophy); group 2, thoracic wall diseases (tumors or costal osteomyelitis); group 3, iatrogenic deformities (acquired Jeune's syndrome, costal graft placement related conditions); and group 4, post-traumatic deformities. Therapeutic procedure is chosen according to the disease etiology. So, group 1 requires treatment of the pathologic process, while group 2 cases are managed by oncology or infectology methods of treatment. In groups 3 and 4, thoracic wall reconstruction is needed. Iatrogenic deformities mostly develop due to poorly performed correction of funnel breast with costal cartilage removal in young patients, with reduction in thorax motility. In case of post-traumatic deformities, pathologic movement as the result of the affected rib pseudoarticulation is frequently observed. Therefore, great caution should be invested to avoid any lesion to the costochondral and costosternal junctions[9].

Conclusion

During the 1985-2005 period, 105 patients with chest wall deformities were operated on at our department by classic method according to Ravitch or a modification according to Welch. In all patients, satisfactory correction of the defect was achieved in early postoperative period and maintained for years. Reoperation for relapse of the disease was required in only two patients. The rate of postoperative complications was somewhat higher than the figures reported in the literature, while the patients' satisfaction with the correction achieved was comparable with literature data. While using original method according to Ravitch, the length of hospital stay was longer than that reported from other centers in the world, whereas the introduction of the method modification reduced it to a mean hospital stay comparable with other centers. The advent of new, less invasive methods of treatment such as Nuss technique, has led to further improvement in some segments of operative treatment, however, classic operation remains the main, reliable and verified treatment modality. The time to come will show which of the methods is superior to the other, although both have some advantages as well as shortcomings.

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Sažetak

NAŠA ISKUSTVA U LIJEČENJU DEFORMITETA STIJENKE PRSNOG KOŠA

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Deformiteti stijenke prsnog koša su relativno rijetke bolesti nepoznate etiologije koje se javljaju u djetinjstvu i adolescenciji. Primjetna je obiteljska pojavnost deformiteta uz vrlo rijetko spontano izlječenje. Kirurško liječenje predstavlja jedinu mogućnost ispravljanja deformiteta bilo klasičnim operacijskim načinom ili minimalno invazivnom metodom liječenja (endoskopskim putem). Među kirurzima nema ujednačenog stava o tome u kojem je životom razdoblju najbolje učiniti ispravljanje deformiteta, jer su rezultati vrlo dobri bez obzira na primijenjenu metodu liječenja. U ovom radu prikazani su rezultati liječenja kod 105 bolesnika operiranih klasičnim načinom između 1985. i 2005. godine u Klinici za torakalnu kirurgiju Kliničke bolnice za plućne bolesti "Jordanovac".

Ključne riječi: *Deformiteti stijenke prsnog koša – patologija; Deformiteti stijenke prsnog koša – kirurgija; Deformiteti stijenke prsnog koša – terapija; Kirurški zahvati operacijski – metode*