

A historical outline of surgical treatment for congenital chest wall deformities

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Abstract

This paper presents the most important events concerning the surgical treatment of congenital chest wall deformities from the early twentieth century to current times. This historical outline features the most deserved and most prominent surgeons in this field, such as F. Sauerbruch, M. Ravitch, D. Nuss, and others. We also recall the Polish pioneers of the surgical treatment of chest wall deformities, including M. Rzepecki, R. Drews, and J. Fibak, at the same time referring to contemporary experiences.

Key words: history of medicine, thoracic surgery.

Streszczenie

W pracy przedstawiono najważniejsze wydarzenia dotyczące leczenia chirurgicznego wrodzonych deformacji ściany klatki piersiowej od początku XX wieku do czasów współczesnych. W rysie historycznym wyszczególniono postaci najbardziej zasłużonych i najwybitniejszych chirurgów, jak F. Sauerbruch, M. Ravitch, D. Nuss i inni. Wiele miejsca poświęcono także polskiemu pionierowi leczenia wad ściany klatki piersiowej, przywołując sylwetki znanych powszechnie chirurgów, takich jak M. Rzepecki, R. Drews, J. Fibak, jednocześnie odwołując się do obecnych doświadczeń.

Słowa kluczowe: historia medycyny, torakochirurgia.

Deformities of the anterior chest wall constitute approximately 90% of all congenital chest defects and occur in 0.01-0.1% of the total population [1]. The first one in terms of incidence, pectus excavatum (hollowed chest), occurs five times more frequently than the second – pectus carinatum, or pigeon chest, which is observed twice as frequently among men as among women. The frequent occurrence of chest wall deformities is explained by genetic determinants, which pertain to 45% of patients; the gene responsible for the occurrence of the deformity is located in the long arm of chromosome 18 [2, 3].

Although no racial predisposition has been found in the incidence of pectus excavatum, it is encountered among patients of Caucasian descent much more often (98%) than among Latinos (9%), Asians (2%), or African-Americans [4, 5].

English literature concerning the defect employs primarily the Latin term pectus excavatum, while German authors often translate it as “*trichterbrust*”. Among the many attempts to unify the related nomenclature, the Greek translation “*chone-chondrosternon*” proposed by A. Ochsner (1896-1981) and M. DeBakey (1908-2008) should be noted, along with *koilosternia*, *thorax en entonnoir*, *pectus infundibuliforme*, or, finally, a term which can still be encountered – *cobbler’s chest* (in Polish terminol-

ogy – “*klatka szewska*”). The above did not, however, gain popular acceptance [6].

The most frequent type of pectus excavatum is a symmetrical deformity consisting in a depression of the lower part of the sternum and the adjoining costal cartilages, which occurs in 45% of cases. The defect may also take the form of an asymmetrical sternal depression with sternal rotation (15%), a shallow and extensive depression of the anterior chest wall (8-22%), or it may be a part of a combined pectus carinatum/excavatum deformity (2%) [7]. Among the less frequent types of the pectus carinatum defect, the forward displacement of the mesosternum, accompanied by bilateral, symmetrical protrusion of costal cartilages, is observed most often [7]. It was termed “*chondrogladiolar*” or “*lower protrusion*” by H. Brodtkin [8]. Asymmetrical deformity with normal sternal position, forward displacement of the costal cartilages on one side, and normal or concave positioning of the cartilages on the other side is less commonly encountered [7]. Finally, the least frequently described form of the defect consists in the protrusion of the episternum with a corresponding depression of the mesosternum or the xiphoid process [7, 8].

Chest wall deformations have been known since the earliest times. Hippocrates described pectus carinatum

as a “sharply pointed chest” and reported that the patients were “affected with difficulty breathing” [9]. The oldest material evidence of the existence of chest wall deformities was provided by the discovery of two cases of sternum excavatum among a set of 48 preserved thoracic skeletons found in Hungary in graves dated between the 10th and 16th centuries C.E. [10].

Recent archeological works conducted under the Ripon Cathedral in Yorkshire, England, uncovered a well-preserved skeleton with signs of pectus carinatum; carbon dating indicated that its owner lived in the 15th century C.E. The skeleton belonged to a young woman, and the positioning of her body in the grave reflected her high social status [11].

Pectus excavatum was first described in 1594 by the French naturalist, botanist, and physician Johann Bauhin (1541-1612). In his work “Sterni cum costis ad interna reflexis nativa spirandi difficultatis causa”, published in the treatise by Johannes Schenck von Grafenberg “Observationum rarum, novarum, admirabilium et monstruosorum” (Frankfurt: Tomus I, Librum II, 1600; 507-508), Bauhin described a case of severe pectus excavatum in a patient who suffered from extensive lung compression, dyspnea, and paroxysmal cough [3].

Hereditary predisposition to pectus excavatum was first noted by W. Coulson in 1820, as he described three brothers with this chest deformity [12]. In 1872, C. Williams wrote about a 17-year-old patient with pectus excavatum, whose father and brother had a similar defect [13]. However, the first to describe pectus excavatum was Eggel in 1870 [7]. Numerous reports of chest deformities were published in the 19th century, including the work of W. Ebstein (1836-1912) from 1882, in which he presented 5 cases of patients whose recommended treatment was fresh air, breathing exercises, aerobic activities, and lying in the lateral position [14, 15].

The first attempt to treat a congenital chest deformity surgically was undertaken in 1911 by L. Meyer; he removed the patient’s ribs 2 and 3, but failed to improve the appearance of the chest [16].

Another attempt was made by Ferdinand Sauerbruch (1875-1951) in 1913. His patient suffered from severe dyspnea at rest and dysrhythmia, which made it impossible for him to work in his father’s watch factory. During the surgery, F. Sauerbruch removed a section of the anterior chest wall within the range of costal cartilages 5 to 9, along with the adjacent sternal segment. After the operation, the patient’s heart could be seen beating under the cover of the muscle flap, but he was able to function and work without dyspnea, and he married 3 years later [16]. In the 1920s, Sauerbruch conducted another corrective surgery employing bilateral costal cartilage resection and sternal osteotomy [2, 16]. He also advocated applying external traction to the sternum in order to hold it in its corrected position for 6 weeks postoperatively. Sauerbruch’s technique was later popularized by other surgeons in Europe and the USA, and, 20 years later, it was used by Mark Ravitch (1910-1989) [16].

In 1939, A. Ochsner and M. DeBaakey published a collection of pictures and descriptions reviewing the various

operative techniques of the time [17]. The high mortality associated with surgery involving extensive chest wall resection led them to advocate less invasive operations. Also in 1939, L. Brown from San Francisco presented treatment results of his two patients and suggested that corrective surgery can be limited to the removal of the ligaments joining the sternum with the diaphragm [18]. The theory concerning the existence of short diaphragmatic ligaments and their pull on the deepest segment of the sternum survived in the field of surgery for the next 60 years, until it was finally disproved by the introduction of thoracoscopic visualization techniques. Nonetheless, M. Ravitch was inspired by L. Brown’s article and proposed more radical sternal mobilization involving the transection of all sternal attachments, including the intercostal neurovascular bundle, rectus abdominis muscle, and attachments of the diaphragm, as well as the excision of the xiphoid process [14].

Throughout the first half of the 20th century, corrective surgery of chest wall deformities was burdened with high perioperative mortality. Out of the 25 patients presented by Ochsner and DeBaakey, few enjoyed any advantages and 6 died as a result of the operation. As late as in 1949, M. Ravitch published a report in which he described a modified Sauerbruch technique used in 8 patients with chest wall deformities. He believed that freeing the sternum of all its attachments would prevent it from sinking further back into the chest, and that pectus excavatum would be eliminated without the need for external traction. The method proposed by Ravitch consisted in the parasternal excision of the most deformed costal cartilages, followed by freeing the sternum and breaking it transversely to the front. The shortened costal cartilages were subsequently reattached to the rim of the correctly positioned sternum by means of non-absorbable sutures [14]. M. Ravitch propagated this technique as late as in 1986.

In 1952, M. Ravitch was the first to conduct pigeon chest surgery by excising numerous deformed cartilages and performing double osteotomy of the sternum [8]. More techniques related to this surgery were described in 1953 by C. Lester. However, the partial excision of the anterior sternal wall was soon abandoned, due to the propensity for heavy bleeding and unsatisfactory results of the correction. Another, less radical surgical method consisted in subperiosteal excision of the sternum [8]. E. Chin (in 1957) and H. Brodtkin (in 1958) advocated the additional transection of the xiphoid process and suturing the rectus abdominis muscle to the upper parts of the mesosternum. However, the method (known as “xyphosternopexy”) often caused the sternum to move backwards, especially in patients with soft, flexible chests [8, 19]. Chin’s method was later modified by R. Howard from Melbourne, who combined subperiosteal excision of costal cartilages with sternal osteotomy [8, 19]. In 1960, M. Ravitch described a different approach to treating pectus carinatum, based on 3 cases. He performed 1- or 2-stage resections of the costal cartilages and then sutured them back in the corrected position. In turn, B. Ramsay treated patients with asymmetrical chests by fill-

ing out the “concavities” with the rectus abdominis muscle [8]. In 1963, F. Robiscek described a method involving subperiosteal excision of cartilages, transverse displacement of the sternum, and excision of the forward-displaced lower segment of the sternum. The rectus abdominis muscle and the xiphoid process were then sutured to the newly formed lower part of the sternum. In cases of asymmetrical deformities, he employed bilateral cartilage resection and double osteotomy of the sternum [8].

In 1953, R. Gross from Harvard published a textbook entitled “The Surgery of Infancy and Childhood”, in which he described the treatment of 8 patients with pectus excavatum. He reported good results of corrective surgery involving cartilage resection in two locations: in the vicinity of the rib attachment and near the sternal attachment, followed by sternal osteotomy. Five years later, K. Welch from the same clinic demonstrated very good results of surgery conducted in 75 patients without the need for transecting the intercostal bundles or the rectus abdominis muscle [20].

Corrective pectus excavatum surgery without the use of external traction was characterized by high recurrence rates. Subsequent operative modifications pertained to the internal stabilization of the sternum. For this purpose, R. Dorner used homologous ribs in 1950, whereas, in 1956, G. Wallgren and M. Sulamaa employed a curved metal bar passed through the lower end of the sternum, which supported the sternum from both sides, keeping it on the ribs [16]. A similar technique was used 5 years later by P. Adkins and B. Blades; it differed in that the bar was not passed through the sternum, but rather behind its posterior surface [21]. The so-called Adkins strut was used routinely in corrective chest wall deformity surgery for nearly 40 years; in spite of the popularity of other, newer operative techniques, it is still employed in many centers. Similar ideas concerning internal support and anterior transverse osteotomy of the sternum were proposed by K. Welch in 1958 [20] and also by A. Haller [22, 23].

Other methods for providing internal support to the sternum employed Dacron vascular graft struts, small titanium struts, seagull wing self-retaining prostheses or substernal mesh bands, or used the bioabsorbable weave technique [16]. Exogenous material was used primarily in adult patients suffering from Marfan syndrome [24]. In turn, plastic surgeons would typically employ silicone implants, which corrected the outward appearance of the chest, but did not eliminate sternal depression [25].

In 1954, J. Judet used a new method, consisting in rotating the sternum completely and fixing it with its dorsal part facing upward [26]. However, this method did not gain popular acceptance outside of Japan, due to its high rate of complications, including infections and necrosis of the sternum [16, 24].

The surgical technique proposed by M. Ravitch, along with its numerous variations described above, became the only efficacious treatment method for chest wall deformities. Some authors noted, however, that costal cartilage resection in children may result in arresting normal

chest development and substituting the costal cartilages by bone or rigid scar tissue, which leads to restrictive limitations and the development of acquired Jeune syndrome (asphyxiating thoracic dysplasia). This phenomenon was described by Pena based on the observation of young rabbits undergoing surgery [27], and was later confirmed by A. Haller in human patients [23]. As a result, many surgeons stopped performing the procedure in small children, waiting for them to pass puberty instead [2]. Damage to the heart and phrenic arteries, and displacement of the corrective bar in the direction of the endocardium, left cardiac ventricle, and abdominal cavity were among the complications noted as occurring after plastic surgery of the anterior chest wall. Meanwhile, the recurrence rate for classical surgery was between 2 and 20.5%, according to various authors [16].

In Poland, corrective surgical treatment of chest wall deformities using a modified Ravitch method was commenced in Poznań by R. Drews (1908-1977) and J. Fibak (1924-2000), who “founded a medical center on Przybyszewskiego Street, to which patients suffering from this defect came from all over Poland” [28]. The technique by Ravitch was here supplemented with external traction by means of the so-called “Fibak frame”, made of wood and strapped to the previously corrected anterior chest wall [29]. This operative method, slightly modified by providing sternal support using a wire, which was fixed to the mentioned “Fibak frame” after closing the wound, was also used for years by the team of M. Rzepecki (1909-1989) in Zakopane [28, 30]. Today, the Ravitch method is still routinely used in treating chest wall deformities by J. Buchwald in Rabka-Zdrój, among others.

Numerous postoperative complications and high rates of recurrence after open surgery caused physicians to search for other, more efficacious and less strenuous operations. In May 1997, at the congress of the American Pediatric Surgical Association (APSA), Donald Nuss presented his 10-year experience with treating pectus excavatum by a method of his design; it consisted in the substernal introduction of an internal corrective bar without the need for costal cartilage resection and sternal osteotomy [31, 32]. The development of this technique was preceded by many years of observations and reflections concerning the Ravitch method. In 1986, D. Nuss reported to have been struck by the flexibility of the rib cartilages – “If these cartilages are so flexible and malleable why am I removing them?” [33]. The minimally invasive Nuss technique soon spread among physicians, and it became the primary surgical treatment for pectus excavatum; widely accepted, safe, and extraordinarily efficacious. The procedure is characterized by short operation time, relative simplicity of performance, low recurrence rate, high efficacy, and permanent corrective results. It is noteworthy that the idea behind the technique was first described in 1925 by Zahradnicek. When treating a 16-year-old boy, he used two metal wires introduced through the sternum, in this way maintaining its traction [17]. More than 60 years had to pass for Zahradnicek’s idea, which was completely dismissed by his contemporaries, to be widely implemented in clinical practice.

To date, many modifications have been introduced to the method originally proposed by D. Nuss, such as the introduction of videothoroscopic control, the stabilization of corrective bars by suturing them to the ribs with non-absorbable sutures and the use of metal supports, the introduction of specialized surgical equipment designed with this method in mind, and the employment of titanium implants, particularly in the case of patients who are allergic to the steel components from which traditional corrective bars are constructed [33].

In Poland, Nuss's method also spread quickly and gained popular acceptance. Its first propagator is J. Dzielicki, who is also in possession of one of the largest operative materials [35]. It is currently performed in most thoracic surgery centers. Since 2002, we have performed surgery on more than 900 patients with various congenital chest defects at the Thoracic Surgery Clinic of Poznań University of Medical Sciences.

Recently, attempts have been made to employ non-invasive methods in the treatment of pectus excavatum, such as the use of vacuum pumps or a magnetic field, consisting in providing external traction to the sternum. However, both techniques have limited application. Employing vacuum pumps is associated with high recurrence rates and damage to the skin, while the sternal traction method, using two magnets facing each other, is still in the phase of clinical tests, but already it seems that it will only be applicable to small deformities. Therefore, surgical treatment still remains the only efficacious method for treating chest wall deformities [36].

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