Pectus Excavatum

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Abstract

Among all chest wall deformities Pectus Excavatum (PE) or funnel chest represents the most common congenital chest wall deformity accounting for 90 % of all deformities. The main characteristic is the depression of sternum and lower cartilages (Langer, Herrn JW Wiener med Zeit 49:515, 1880) with an incidence between 1 and 8 per 1000 children.

Keywords

Pectus Excavatum • Funnel chest • Congenital chest wall deformity

Pectus excavatum (PE) or funnel chest represents the most common congenital chest wall deformity accounting for 90 % of all deformities. Main characteristic is the depression of sternum and lower cartilages (Fig. 4.1, 4.2, and 4.3). The first description came from Bauhinus [1] in the sixteenth century. Another documented description of an appearance of the thorax could be found in 1860 by Woillez [2] and in 1863, von Luschka [3] reported about a 6-cm deep depression in the thorax wall of a 24-year-old man. Eggel [4] in 1870 published the first comprehensive case report of a patient with a funnel-formed thorax depression calling it a 'miraculum naturae'. He assumed that the reason for the deformity would be a weakness and an abnormal flexibility of the sternum caused by nutritional disturbance or by developmental failure. Individual case reports followed by Williams [5], Flesch [6] and Hagmann [7]. The latter believed that overgrowth of the ribs causes the depression of the chest. Langer and Zuckerkandel [8] favoured the hypothesis of a developmental failure, taking place in utero, in which the lower jaw of the foetus is responsible for the deformity by pushing on the sternum as a result of too high intrauterine pressure. Meyer performed the first operation of PE in 1911 with the removal of the rib cartilage [9]. He also analysed the removed cartilage microscopically and identified an unspecific degeneration.

The incidence of PE has a ratio between 1 and 8 per 1000 children [10]. Interestingly, males are more often affected, with a gender distribution

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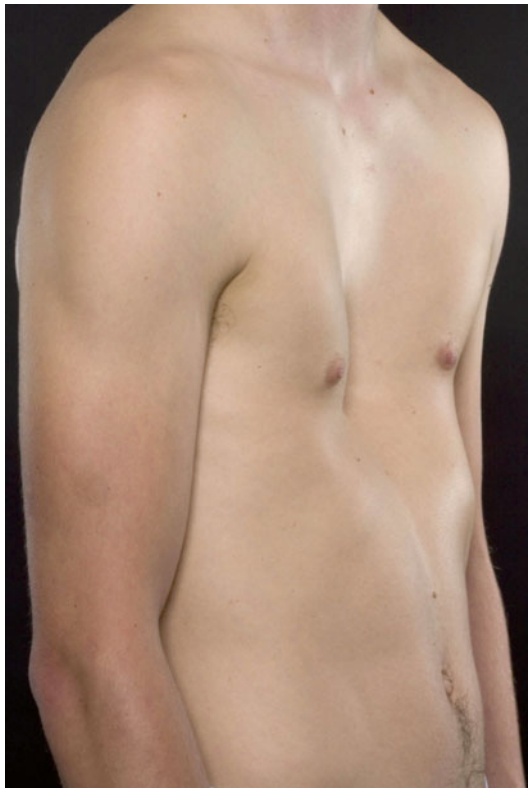

Fig. 4.1 Right side view in a 16-year-old

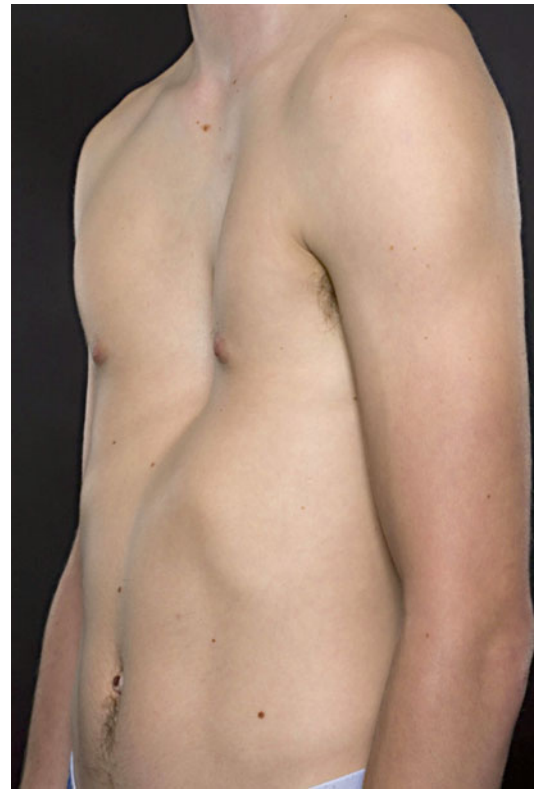

Fig. 4.2 Left side view in a 16-year-old

between 2:1 and 9:1 [11]. From the 19th century it has been recognised a genetic predisposition since a positive family history could be found in up to 43 % of PE cases [12, 13]. However, a specific genetic defect has not yet been found. Numerous syndromes are associated with PE and have been well described [14] where connective tissue disorder occurs in less than 1 % of all cases [15].

The majority of the patients with PE are tall, slim with associated scoliosis [14–16]. Severe depression of the sternum can cause displacement of the heart and reduction of lung volume [13, 17]. As a result of the anatomical changes, chest pain [12, 13, 15], fatigue [15], dyspnoea on exertion [12, 13, 15, 18], respiratory infections [13], asthma symptoms [13], palpitations [12] or heart murmurs could occur [13]. Several cases with mitral valve prolapse [13, 15, 19], mitral valve regurgitation and ventricle compression could be found [15, 17, 19]. For the latter, Coln [19] demonstrated that 95 % of 123 patients had cardiac compression. Even a single case report of syncopal symptoms has been reported. The pulmonary and cardiovascular functions of patients with PE deformities were analysed in many investigations and have revealed measurable deficiencies [12]. Fonkalsrud [13] reported that the symptoms of many untreated PE patients become progressively worse with age and he recommended an operational intervention for both young and adult patients.

In contrast to these descriptions of more or less severe clinical signs, symptoms affecting daily life activities are either rare [20]. Therefore, some authors described the indication for a PE correction to be primarily cosmetic.

Numerous clinical studies described an improvement of pulmonary and/or cardiovascular symptoms and improvement in the subjective well-being after surgical correction [9, 13, 16–18]. Malek [18, 21] concluded that an operative intervention improves cardiovascular but not pulmonary function. Guntheroth [22] and Spiers as well as Johnson [23] re-evaluated the source data of Malek's meta-analyses and stated that due to

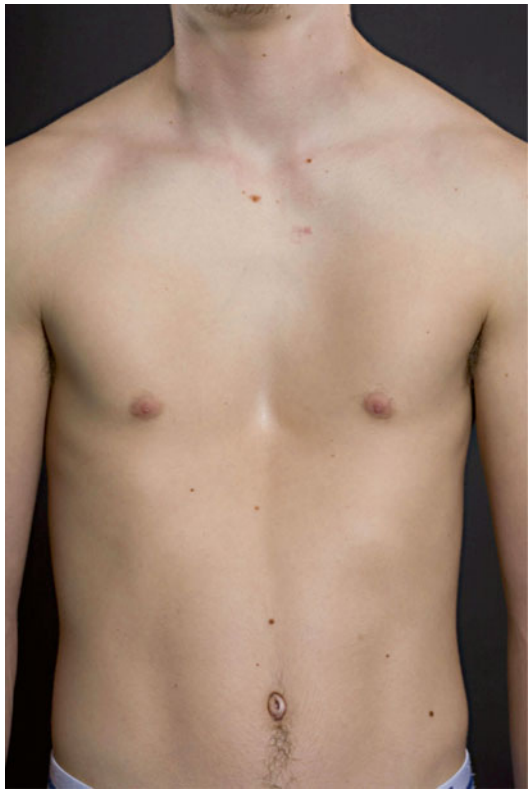

Fig. 4.3 Front view in a 16-year-old

relevant methodological deficits, these data failed to demonstrate any improvement of cardiac function. In this context, Aronson et al. [24] could not show an improvement in lung function parameters after Nuss procedure. Regardless the impact of the chest wall surgery to cardiopulmonary function the effect to psychological status of the patient is significant [15]. Numerous studies confirm that deformities cause relevant social discrimination, especially during adolescence, leading to the socio-psychologic problems [15]. A multicentre study demonstrated that the surgical repair of PE patients improves these socio-psychologic problems. [25–32]

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