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Diagnosis, Therapy and Current Developments

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Preface

Several publications, case reports, reviews, and new surgical techniques, either as modifications or new developments of the pectus excavatum or carinatum surgery have inundated the literature databases over the past decade. The shear quantity of individual publications on the most varied technologies impedes the appropriate flow of information from the primary therapist to the patient and his or her parents, and sometimes is even confusing for the physician or surgeon himself/herself. To date, no comprehensive work with an overview of the current surgical, comparatively rare or non-surgical alternative treatment possibilities is available. Therefore, with this interdisciplinary work we made it our task, while making no claims to completeness, to create an overview of diagnostic measures, therapeutic options and the follow-up treatment of congenital thoracic wall deformities.

This work is dedicated to all the authors for their valuable and precious contributions to the further development and promotion of diagnostic and therapeutic options in the treatment of patients suffering from such strains.

We are also exceptionally grateful to the photographers Karin Langert and Angelika Feichter for their distinctive art and high-quality photo documentation.

Equally we have to express our thanks to Professor Werner Jaschke and his team from the Department of Radiology, Medical University Innsbruck for the generation of sophisticated radiologic imaging and their readiness to provide us with this artwork that enriches the book so enormously.

Without this comprehensive support this book would definitely not have its appealing vividness.

Anton H. Schwabegger, MD, MSc

Foreword

During the first decade of the 21st century, it is highly educational to look back and observe the progress that was made in thoracic surgery during the 20th century. In fact, the 20th century could be called as the Century of thoracic Surgery! Ira Rutkow in his book: Surgery: An Illustrative History states "through the last decades of the nineteenth century, operative intervention in the heart, lungs and other organs in the mediastinum and thoracic cage usually had fatal results. Accordingly, little interest was expressed in the establishment of such surgery as a specialty. As more and more papers on thoracic surgery were being presented during the first 20 years of the 20th century, however, this attitude changed".

Even at the beginning of the 20th century, anesthesia was administered by face-mask and the lungs would collapse as soon as the chest was opened. It is not surprising, therefore, that early attempts at chest wall reconstruction were designed to approach the problem externally to avoid opening the pleural cavity. Sauerbruch even went further to design a differential negative pressure chamber, which encompassed the patient's body from the neck down and was large enough to admit the surgeon as well. According to Meade in his book: A History of thoracic Surgery, "it was not until after the first World War that Rowbotham and Magill used a simple wide bored rubber tube inserted into the upper trachea. A cuffed endotracheal tube did not become available until 1932." This finally allowed thoracic surgery to expand to the point of permitting wide resection of chest wall structures, lung resections, and later even open-heart surgery. However even during the 1950 polio epidemics, patients who developed respiratory paralysis were placed in "iron lungs" not unlike Sauerbruch's differential pressure chambers. then in the 1960s, ventilators were developed which opened up the era of "Intensive Care Units" and ever-larger operations on ever-sicker patients up to and including heart–lung transplants. Wide resection of the anterior wall structure for the treatment of chest wall deformities was advocated by all major medical centers – even in very young children. In the 1980s, there was a huge paradigm shift with the introduction of fiber optics into the field of surgery. Suddenly the emphasis changed from ever-larger incisions to ever-smaller incisions starting with 10-mm thoracoscopes down to 2-mm thoracoscopes. It is with this historical background in mind that this textbook reviews the present day management of congenital and acquired thoracic wall deformities. the management of the two most common chest wall anomalies – pectus excavatum and carinatum – have undergone equally dramatic paradiagm shifts from wide or radical resection of anterior chest wall structures to minimally invasive procedures and even non-surgical approaches including suction cups and pressure braces.

As recently as 1990, anterior chest wall surgery was considered to have matured with no new innovations. Suddenly this has become an exciting and dynamic area of surgery with new ideas and innovations being prescribed at conferences and in medical journals almost on a monthly basis.

Donald Nuss, MB, ChB

Contents

Introduction

Anton H. Schwabegger

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Congenital wall defects, if no functional deficit by cardiac or pulmonary impairment exists in marked deformities, predominately encumber patients because of their unsightly aesthetically unpleasant stain. The appearance of such a deformity is not concealable unclothed and even clothed in the keel chest deformity may not be camouflaged due to its prominent bulge. During not only puberty but also during adolescence and later such deformities lead to shun behaviour and social retreat.

Not only for leisure-time activities, which are increasingly focused on life-style and body constitution, but also for the process of partnership initiation such a deformity stain represents a significant social handicap. The more seldom, true cardiopulmonal problems, caused by deep thoracic wall depressions with displacement or even compression of heart and lungs, usually are an exception for indication setting to thoracoplasty. In most cases however, the need for correction is based on evident social adaptive difficulties and impairment of worth living sense.

In former times and even a few decades ago thoracic deformities without or with only minor functional impairment were settled as a simple non-aesthetic stains, not at all considering the need for correction, thus they were scarcely corrected by surgical interventions. Herewith the psychic state of derangement of affected patients was not adequately considered or even neglected.

Potentially because of ignorance or lack of knowledge about therapeutic options and thus embarrassment, inappropriate medical counsels or advices were given to the patient like "it will resolve by time and body growth", "one can camouflage it by clothing", "muscle training cures the deformity" or "surgery is much too risky".

Nowadays patients and parents, usually by means of electronic media, are much better informed about the available therapeutic options, occasionally even more in detail than some physicians and thus are much more demanding for correction of their unpleasant stain. Furthermore all available minor or major invasive methods of surgical and alternative supportive actions are more developed by technical and surgical methods nowadays. Increasingly minor deformities are requested to be corrected as rather aesthetic interventions.

Since the publication of Donald Nuss about the success of the minimally invasive repair of pectus excavatum (MIRPE) in 1998 the demand for correction of all sorts of thoracic wall deformities boomed almost all over the world. The procession of the minimally invasive pectus-bar, a modification of formerly more invasive surgical methods is still ongoing. This abovementioned publication by Nuss was a report of experiences predominately in children, but after a follow-up of 10 years though, it was very well suitable for reliable evaluation. Nevertheless in many institutions this report gave rise to a euphoric application to all sorts of funnel chest deformities and even up to late adulthood. It seemed to develop into a method of correction "for all seasons". Ensuing to that a myriad of publications appeared in the medical literature databases. However, most of these quite early consecutive publications just described more or less small series of application and merely clinical observations without adequate or inadequate follow-up. There were only a few reports about the evaluation of distinct long-term results in different ages and genders or potential intricate complications. That is why, caused by euphoric presentation through all kinds of media in many places at times led to uncritical employment for all kinds of deformities even in aged adults with already rigid thoracic cages. The following and subsequent failures and rebounds in particular cases, although rarely published, induced the development of modified and combined techniques or led to reminiscence to established older methods.

However, the development is still going on and the MIRPE focuses on a defined but broad entity of funnel chest expressions and indications, whereas the ongoing discussion and confrontation with alternative methods defined special indications for the application of other available and occasionally more suitable techniques in children, adolescents and adults as well.

Nevertheless, many of the techniques described in the following chapters should be performed only at specialized centres, which fulfil the requirements of broad experience by sufficient numbers of cases treated. On the basis of the complexity of a broad variety of expressions of anterior thoracic wall deformities the selection of an appropriate treatment procedure for the medical requirements and increasingly individual aesthetical claims requires much experience and mostly an interdisciplinary access and discourse, to offer the patient not only the best but also the correct procedure in every individual case. Thus adequate know-how about selective indication setting and skill for standard techniques equivalent to alternative methods may enhance the overall quality of selective treatment. Furthermore interdisciplinary access to an intensive care unit at any time and close relationship to thoracic surgeons for the unintended case of severe potential complications conjoined to the invasive methods of correction must be available.

This book was initiated for the very reason of creating a wide-ranging survey over diagnostics and a multitude of different surgical treatment options for congenital thoracic wall deformities. It was also an endeavor to join established treatment procedures and expertise from abroad as well as alternative, even non-surgical interventions to provide with sufficient information for physicians in advance or during first consultations of patients afflicted with such congenital thoracic wall deformities.

Deformities of the anterior thoracic wall

2

2.1 Functional anatomy of the thoracic cage

Bernhard Moriggl

2.1.1 Introduction

The individual shape of the *thoracic cage* in both men and women has considerable impact regarding phenotype and expression of human beings of different races. It is thus not surprising that the thorax finds inherent and manifold mention in linguistic usage of all peoples (e.g., "chest out!"; "up to one's chest"; and so forth). It follows that any noteworthy deviation from a considered norm may lead to mental trauma and/or psychic problems. Although obviously not the major problem, functional impairment may occur, not least justifying for sketching the basic *Functional anatomy* (Sections 2.1.2 and 2.1.3).

> "*Function without morphology seems somewhat unearthly; morphology without function is dead!*" (comment B. M.: going to bat for a "lively Anatomy").

In brief and with respect to operative procedures, relevant *Topographical anatomy* (Section 2.1.4) has also to be covered.

> "*Surgeons have more and more come to realize that knowledge of* (comment B. M.: topographical) *anatomy in surgical procedures is the best safeguard to avoid surgical injuries*"
>
> *Nicholas A. Michels*

The attempt of this chapter "*Functional anatomy of the thoracic cage*" is to give an idea of selected anatomical concepts related to this entity that surgeons might find useful. The author is well *aware* of the fact that *some* of the given descriptions and *interpretations* are *not* in full accordance with *general beliefs*. This is at first a result of extensive experience in applied anatomy gained during professional education and training in the field, and secondly of fruitful discussions as well as cooperation with my clinical colleagues of various (surgical) disciplines I have been fortunate enough to experience over more than 22 years now. It is hoped that the contribution will also encourage the reader to refresh, go back or – even better – deepen aspects of "*lively Anatomy*" that are of further value but have not been dealt with exhaustively.

> "*Once you start studying Medicine* (comment B. M.: and so with Anatomy) *you never get through with it.*"
>
> *Dr. Charles Horace Mayo*

2.1.2 Developmental aspects

2.1.2.1 Ribs and sternum (see Figs. 1 and 2)

It is crucial to understand that the thoracic skeleton anlagen derive from the same blastema. All anlagen of the ribs' ventral end will fuse longitudinally to form the cartilaginous paired sternal bars (bands, plates). The latter gradually move toward the midline pushed by elongation of the ribs. Finally the two strips blend zip-like in a craniocaudal direction to form *the sternum*. Developmental disturbances of that process may result in a cleft sternum. Within the cartilaginous primordium of the sternum the first ossification centers occur in the manubrium, others in the body of sternum follow. Usually those in the upper part are single (as in the manubrium), while the others are paired but often asymmetrically arranged (Fig. 1). In clear contrast to their appearance, union of ossification centers in the body of sternum proceeds from below upward. Incomplete fusion of paired centers in the lower third of the bone results in the well-known variation of a sternal foramen (Fissura sterni congenita). As a rule, synostosis of centers for the body starts with puberty and is finalized between age 20 and 25. Bone development within the xiphiod process is postponed compared to the rest of the sternum; in some sterna this part stays cartilaginous. As a whole, number and location of centers of ossification in the sternum varies considerably as this is related to completeness and time of fusion of the above-mentioned sternal bars. Bone formation within the flattened bars of cartilaginous *ribs* starts posterior near the angle, moves ventrally but comes to a sudden stop in this direction (as early as the 4 month of intrauterine life), the reason for existence of the functionally most significant costal cartilages (Fig. 2).

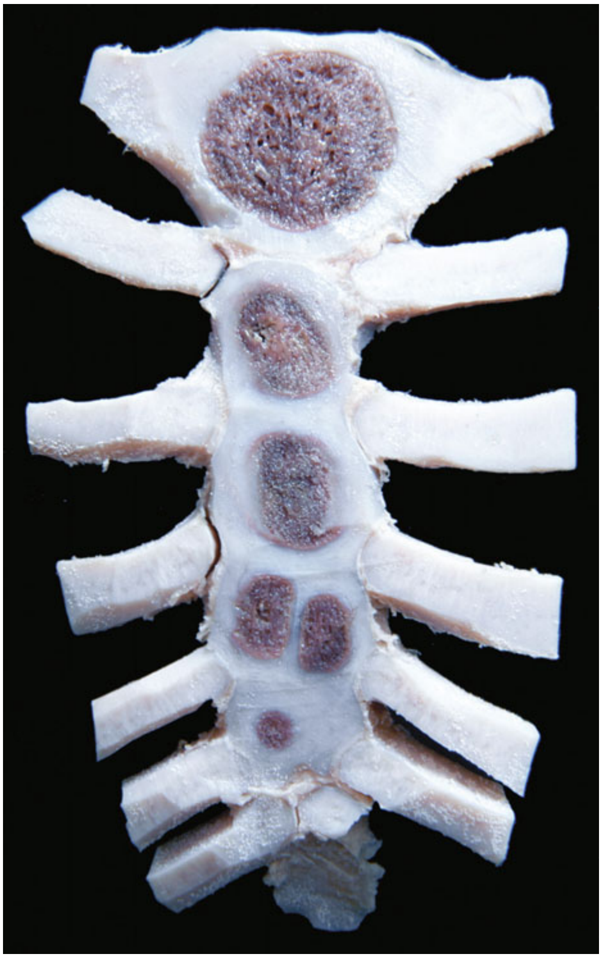

Fig. 1. Development of sternum. Cartilaginous sternum of a baby: note unpaired (upper half of body) and paired (lower half) ossification centers within. The latter are often asymmetrically located and do not appear synchronized!

These few glimpses make it understandable why *irregular growth* of costal cartilage, deviant *ossification* in the (lower part of) sternum, or both, may contribute to the formation of funnel as well as keel chests.

2.1.2.2 Thoracic spine

As for the whole axial skeleton, the thoracic part needs existence of and proper induction by the non-segmental notochord (*Chorda dorsalis*), segmentation and rearrangement by sclerotomes – the mesenchymatous models of vertebrae – and their cartilaginous transformation (first three stages of spine development; for details see textbooks of Embryology). In a fourth and *final step*, ossification starts with the appearance of three primary centers: two in the pedicles of vertebral arches (perichondral), one is endochondral and located

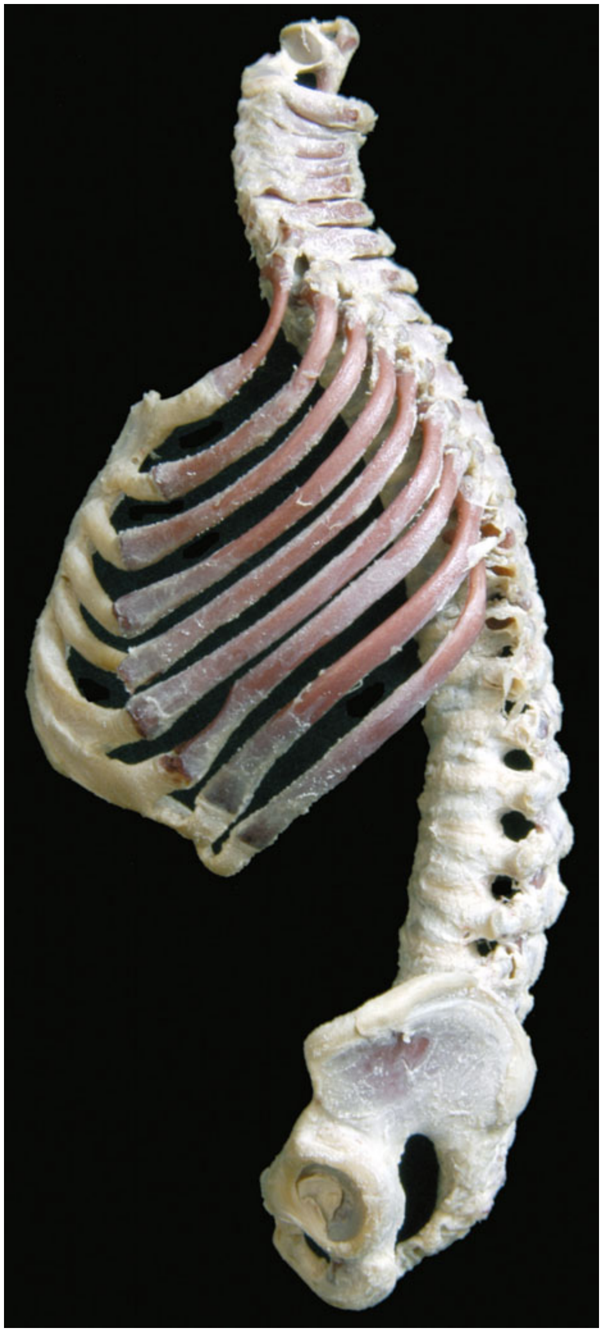

Fig. 2. Spine and left half of thoracic cage in a newborn. Starting from posterior, bony transformation (in red) of the ribs stops in considerable distance to the sternum to leave the cartilaginous part of each rib (cream-colored) and guarantee formation of the costal arch

in the vertebral body. Osseous fusion to a single vertebral arch occurs in the first year of life, synostosis with the body starts in the third. Out of the numerous secondary ossification centers, those in the epiphysis

of the vertebral body are of practical value (for growth, form, and stability of the vertebra!). Here, approximately with the age of 8, bone formation is in a ring-like fashion ("annular epiphysis") for the circumferential part of both vertebral body surfaces. They start fusing with the rest of the body about the age of 18. Under normal circumstances, with the age of 25 all ossification processes in the thoracic spine should have come to a complete stop. Practically speaking, the final shape as well as full load-bearing capacity of the spine is achieved considerably late!
Because the thoracic part of the vertebral column is both, *morphological and mechanical basis of the thorax*, this has to be considered with any exceptional loading or (surgically) applied structural changes during childhood and adolescence, respectively.

2.1.3 Mobility and motion

2.1.3.1 Bones, joints, and the influence of anthropological factors (see Figs. 3 and 4)

The shape of the chest as such does not noticeably influence function of thoracic viscera. The all-decisive factor remains *mobility*. That is why usually only severe forms of chest (wall) deformities are accompanied by respiratory or circulatory problems.
Strictly speaking the skeletal basis of the thoracic cage would only include both, *ribs* and *sternum*. From a functional point of view however, the *thoracic spine* has to be mentioned simultaneously and viewed as a functional unit (see above and Section 2.1.3.3).
The *skeleton* of the chest is an *osseo-cartilaginous framework* containing and protecting essential organs of circulation and respiration. For respiration, mobility of the ribs is the basic prerequisite. This is primarily realized by both, diarthrosis (synovial joints) and synarthrosis (synchondroses in particular). Often disregarded however, the *bony parts* of the *ribs* themselves undergo distortion and thus contribute to the overall *elasticity* of the thoracic cage. To a minor degree, an increase or decrease of thoracic kyphosis plays a role during respiratory movements.
The *costovertebral joints*, both at the bodies, joints of the head of the ribs, and transverse processes, costo-transverse joints at the ribs' tubercles, are diarthrosis with their synovial capsules surrounded by a rather strong ligamentous apparatus (reinforcement and guidance). In addition, the joints of the heads of the ribs (with the exception of 1st, 11th, and 12th) are double-chambered through intra-articular costal ligaments that connect

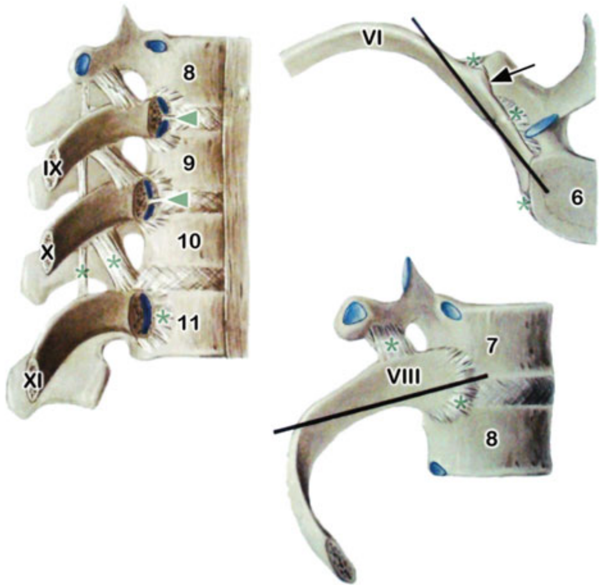

Fig. 3. Costovertebral joints and axis of movements. All costovertebral diarthrosis are reinforced by a strong ligamentous apparatus (green asterisks). The joints of the head of the ribs down to the 10th rib (X) appear double-chambered through additional, intra-articular costal ligaments (green arrowheads). Note flattened articulating surfaces of sixth costo-transverse joint (black arrow). Black lines indicate axis of movement. 6–11 thoracic vertebrae; VI, VII–XI ribs (© Lanz T v, Wachsmuth W, Praktische Anatomie. Rücken. Springer, Berlin Heidelberg, Special Edition 2004, p 41, Figs. 56, 57)

Fig. 4. Sternocostal joints (cartilage in blue). The first junction is always a synchondrosis (blue asterisk), while second (shown here) through fifth are synovial joints; green arrowhead indicates intra-articular sternocostal ligament that regularly creates a double joint cavity at this level; blue arrowhead: manubriosternal synchondrosis; green asterisks indicate strengthening ligaments; I and II ribs; *MS* manubrium of sternum; *BS* body of sternum; *CL* clavicle (© Tillmann B, Atlas der Anatomie des Menschen. Mit Muskeltrainer. Springer, Berlin Heidelberg, 2005, p 397, Fig. 6.12)

the intervertebral disks to the crest of the head of the ribs (Fig. 3). Apart from this ligamentous restriction, differing mobility of named synovial joints also depends on the shape of articulating facets of the costo-transverse joints: the facets are concave in second through fifth vertebrae, allowing for *rotatory motion about the neck* of these ribs, while there are simple *sliding movements* for the 1st rib and 6th–10th, respectively (facets flattened!). The axis of movement runs within the neck of each rib (Fig. 3). It follows that the orientation of this axis (principally directed backward) changes from slightly upward (at the first rib) over transverse to downward (starting from fifth). From a functional point of view it is noteworthy that described morphological peculiarities of facets at the vertebral column (vertebrae 1–7 in particular as ribs 8–10 are only indirectly connected to the sternum) find their parallel in the *Sternocostal joints*: the first of these is always a *synchondrosis*, while second through fifth are *diarthroses* with tough strengthening ligaments. Among the synovial joints, only the second will regularly show an intra-articular sternocostal ligament creating double joint cavities (Fig. 4). The junctions between ribs 6–7 and the sternum are again cartilaginous in most individuals; however, joint cavities may occur. Finally, synovial joints can be found between sixth and ninth costal cartilages, the *interchondral diarthrosis* (almost constant between sixth and seventh).

The thorax as a whole varies as to dimensions and proportions. As in most other parts of the skeleton and apart from individual nuances, sex differences occur: in the female the capacity is less, the sternum is shorter (relatively) and the thoracic inlet is more oblique. Moreover, there is generally greater movement in the upper ribs as in the male, permitting comparatively greater expansion of the upper part of the thoracic cage. Concerning constitution as well as racial characteristics it may briefly be stated that the thorax shares in given general proportions (e.g., typically tall and thin in the leptosome or in distinct tribes).

Among anthropological factors however, and especially in view of functional consequences, the influence of *age* is outstanding. As a rule, changes and adaptation in shape of the framework is easily achieved as long as *elasticity* remains intact. Likewise, the relatively small transverse diameter of the thorax in newborns will gradually adapt to adult proportions with the child's ability to walk (this also partly reflects the influence of muscles and their function; see Section 2.1.3.2). The elasticity of the thorax also guarantees for great resistance to stress (not least operations!). Although the main factor for the essential distortions of the thorax during respiration is elasticity of the *costal cartilages* one should not underestimate the influence of the joints involved. Thus, in addition to the well-known and early occurrence of cartilage *calcification*, *disorders* of *junctions* may well contribute to reduced thoracic plasticity and function. Plasticity and elasticity of the thoracic cage will also be reduced in very *athletic* adolescents. This directly leads to the (functional) role of musculature.

2.1.3.2 Musculature (see Figs. 5–7)

Regularly, when talking about "thoracic musculature", we think of muscles that are primarily concerned in the movement of ribs, meaning with respiration in a closer sense. Nonetheless it is crucial to understand that *all muscles* that are *attached to the thoracic skeleton* have to be considered as "*moving forces*" *of the thorax* in general

Fig. 5. Muscular suspension of thorax and muscles of thorax proper. The scalene muscles (SMm and insert) are attached to vestigial ribs (part of transverse processes) of the cervical spine; together with the sternocleidomastoid muscle (*SCM*) they are also active in inspiration; in the third ICS part of the external intercostal muscles (*EICM*) are fenestrated to show the internal intercostals (IICM; note their fiber orientation!); as the EICM end at the costal cartilages (continued only by the external intercostal membrane) the IICM represent the superficial muscular layer until the sternum (*S*) and are known as "intercartilaginous muscles" (© Tillmann B, Atlas der Anatomie des Menschen. Mit Muskeltrainer. Springer, Berlin Heidelberg, 2005, p 204, Fig. 4.30)

Fig. 6. Thoracic shield viewed from inside: muscles of thorax proper; transversus thoracis muscle (*TTM*). Typical appearance of a TTM* (= sternocostalis) and its relationships; *MS* manubrium of sternum; *BS* body of sternum; I and II, VI and VII ribs; *ICIM* intercostales intimi muscles (innermost part of the IICM); note shape and asymmetry of TTMs of both sides; *TAM* transversus abdominal muscle; *DI* diaphragm (note most medially seen slender slip of sternal part) (© Tillmann B, Atlas der Anatomie des Menschen. Mit Muskeltrainer. Springer, Berlin Heidelberg, 2005, p 205, Fig. 4.31)

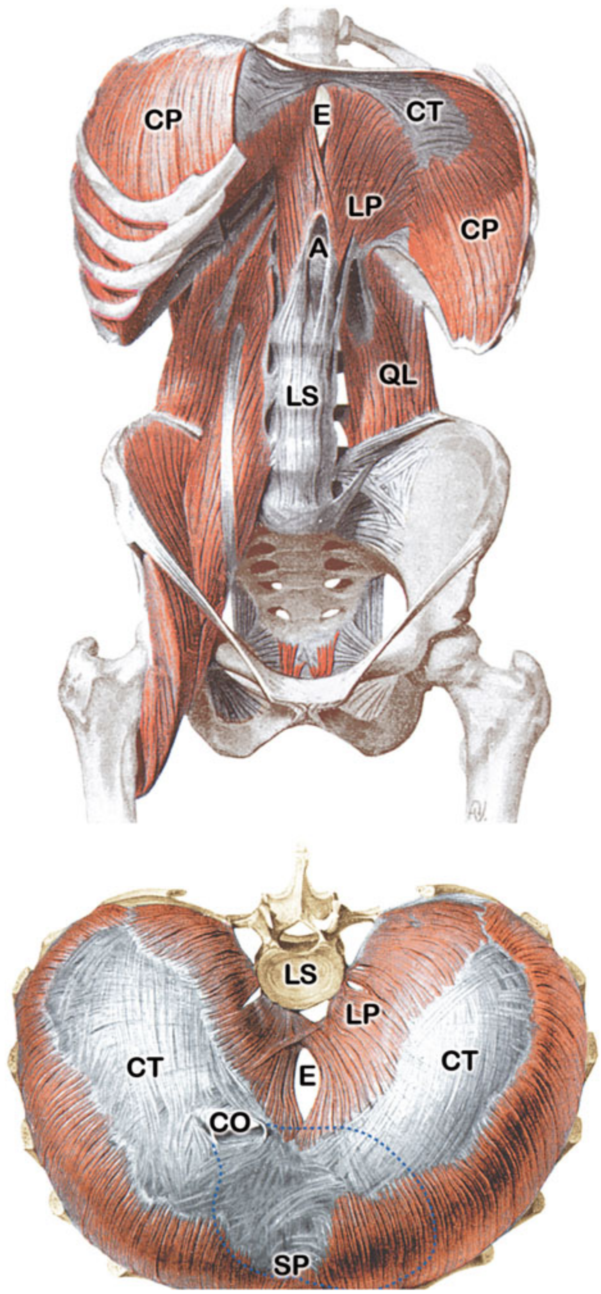

Fig. 7. Dome-shaped diaphragm viewed from anterior and superior. In upper image sternal parts (*SP*) completely, costal part (*CP*) of left side partly removed; *LS* lumbar spine; *LP* lumbar part with crura, esophageal (*E*) and aortic (*A*) hiatuses; *CT*, central tendon with caval opening (*CO*); *QL* quadratus lumborum muscle (© Tillmann B, Atlas der Anatomie des Menschen. Mit Muskeltrainer. Springer, Berlin Heidelberg, 2005, p 212, Fig. 4.40b)

(not necessarily related to breathing!). That way, one has to include parts of the erector spinae, the scalene muscles, migrant ventrolateral muscles (especially the posterior inferior serratus), muscles of the shoulder girdle, the latissimus dorsi and all of those forming the abdominal wall. Especially the latter create a floating balance of forces with diaphragmatic movements during respiration (by synergism and antagonism).

Muscular suspension of thorax

The muscular suspension of thorax to the skull and cervical spine is mainly based on *scalene* and *sternocleidomastoid muscles* of both sides (Fig. 5). They are able to withstand caudally directed traction; prerequisite for them to get effectively involved in inspiration (by lifting the upper part of thorax) is fixation of origins that allow for bilateral and synchronized action. Often underestimated, the *scalene muscles* are most significant for quiet breathing (with increased vigor when head bent backward). The scalene muscles may be viewed as a cranial expansion of intercostals (attached to vestigial ribs of the cervical spine!).

Muscles of thorax proper

Among the muscles of thorax proper, the *transversus thoracis* (better named *sternocostalis*) is probably the least known despite its functional significance (expiratory muscle). In addition, it is of remarkable topographical interest (see Section 2.1.4.1). The official name is misleading in so far as it is a fan shaped muscle (radiating outward from the lower part of the posterior surface of the sternum). The highest fibers run very steep to reach the second costal cartilage (inner surface), the intermediate ones are oblique, while only the lower fibers are transversely orientated (toward the ribs 6–7; Fig. 6). Therefore and importantly, the lowest part of this muscle is contiguous with the highest slips of the transversus abdominis muscle, which in turn interdigitates with the thoracic origins of the diaphragm! Finally, this muscle is often asymmetrical between opposite sides of the same individual (Fig. 6!) and varies considerably in its attachments as well as strength in different people.

The *intercostals* are three superimposed thin muscular layers. Because the *external intercostals* only occupy the spaces between bony parts of the ribs the *internal intercostals* build the superficial layer between costal cartilages (Figs. 5 and 6). The latter part is topographically and functionally referred to as intercartilaginous muscles (active during forced inspiration in contrast to its "interosseous" portion). The innermost part of the internals is separated off as *intercostales intimi*, thus representing the third layer (see also Section 2.1.4). Fiber orientation of the inner two muscle plates coincides and is opposite and nearly at right angles to that of the external muscles, meaning from infero-posterior to supero-anterior. This principally explains their overall antagonism during respiration (externals "in", internals "ex") although opinions toward their functional significance are still not unanimous at all! Despite mentioned controversies it is justified to state that (1) the main activities of all intercostals are in forced breathing! and (2) they act as bracing system for the intercostal spaces (elastic supports).

The diaphragm (Figs. 6 and 7)

This *essential muscle of respiration* is dome-shaped with a peripheral fleshy part arising from all skeletal elements that form the thoracic outlet and from the lumbar vertebrae as well as intervertebral disks (parts of crura of diaphragm; Fig. 7). All muscle fibers converge into the central tendon. It is interesting to note that shortness of this aponeurotic part together with exceptional muscular tension is believed to be one of the etiologic factors for the development of funnel chests. In accordance with this, the deformity is less obvious at birth but becomes increasingly marked with the growth of the individual.

Shape and position of the diaphragm in the thorax (and thus function) depends on three main factors: *posture*, *pressure* from the abdomen (due to both viscera and abdominal muscles) and, most obvious, status of *respiration* itself. The latter is best expressed by the fact that during expiration fibers of the sternal part ascend (Fig. 6), while in maximum inspiration they even descend to the central tendon! To a lesser degree the almost vertically orientated course of costal part and crus bundles (Figs. 6 and 7) will flatten out.

2.1.3.3 The thorax as functional unit, mechanics of respiration

The movements of respiration (more general: forces that move the thorax; see above) can only be fully appreciated if all elements of the thorax (Sections 2.1.3.1 and 2.1.3.2) are viewed as a functional unit. All thoracic *movements result* from a *vast number of single movements*. In addition to autochthonous parts, structures not strictly part of the framework itself have remarkable impact in the *mechanics* (dynamics and kinematics) of respiration.

The mechanic principal of respiration is an alternate increase and decrease of capacity of the thoracic cavity. While in quiet respiration the former requires muscular input, the latter is largely passive.

Increase in volume is based on two processes: (1) with the *active elevation of ribs* both transverse and sagittal dimensions of the thorax are increased (2) the vertical dimension is increased by *contraction* (i.e., *sinking*) of the *diaphragm*.

Elevation of the ribs (and sternum) is possible by rotation around the oblique, antero-posterior axis through their necks (see Fig. 3). The plasticity of costal cartilages allows for their marked torsion during that movement. The scalene and sternocleidomastoid muscles have important functions during inspiration: together with the diaphragm (see below) they are the main muscles concerned in quiet breathing; on the other hand they have important auxiliary function for more accentuated lifting of the ribs by virtue of fixation of the thoracic inlet! With deep inspiration, additional muscles come into action. The external intercostals and intercartilaginous parts of internals become more and more active in an increasing number of intercostal spaces.

Moreover, the greater *descent* of the *diaphragm* will increasingly press on abdominal contents that may lower only by simultaneous relaxation of the superficial abdominal muscles! Toward the end of the diaphragmatic descent, abdominal viscera – above all the liver – will provide enough resistance for the central tendon so that the fibers will elevate the lower ribs and additionally increase diameter.

At the same time however, there is tendency of the diaphragm to pull the lower ribs inward. This is made impossible by simultaneous stabilization through action of the posterior inferior serratus. Another important contributor to effectiveness of diaphragmatic power is the quadratus lumborum by fixation of the 12th rib. Finally, activity of the erector spinae is augmented (influencing curvature of the thoracic spine) and muscles connecting the trunk to upper limbs, pectoralis major in particular, may get involved in forced inspiration (provided the extremities are fixed).

Decrease in volume is guaranteed by the *recoil* of the chest wall and lungs in breathing at rest. For forced expiration (especially against resistance) the muscles of the abdominal wall together with the latissimus dorsi provide appreciable "*external power*".

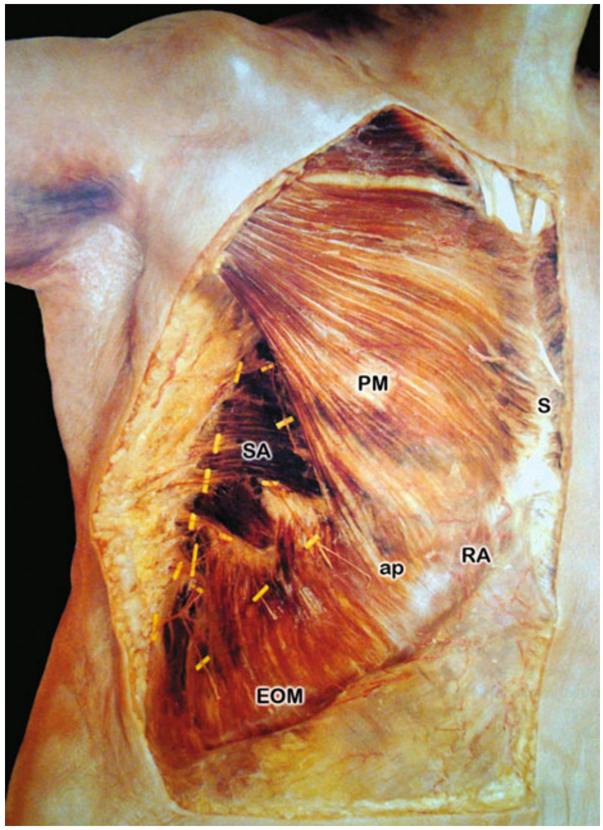

Fig. 8. Muscular blanket of anterior (and lateral) thoracic wall. Both pectoralis major muscles (*PM*) almost completely cover the sternum (*S*) or sternal membrane; *RA* rectus abdominis muscle (transparent through rectus sheath!); *ap* abdominal part of PM; serratus anterior (*SA*) and external oblique (*EOM*) muscles intermingling; emerging lateral cutaneous branches of intercostal nerves lined yellow (© Thiel W, Photographischer Atlas der Praktischen Anatomie II. Hals, Kopf, Rücken, Brust, Obere Extremität. Springer, Berlin Heidelberg, 1999, p 160, Fig. 80)

2.1.4 Pearls of topographical anatomy

2.1.4.1 Anterior thoracic wall (see Figs. 8–14)

Before approaching the anterior thoracic wall, one has to traverse the *pectoralis major muscle*. It is macroscopically characterized by rather big muscle fascicules (or bundles) with considerable amount of loose connective tissue in between. This facilitates "muscle splitting" (as should be done whenever possible instead of detachment leading to prolonged postoperative impairment). The two muscles of either side almost touch each other at the midline, so most of the sternum has a muscular covering (Fig. 8). Short tendons contribute to forming the sternal membrane. Antero-caudally the *sternocostal part* of pectoralis major interdigitates with costal origins of the *rectus abdominis* muscle – noteworthy another essential part of the *muscular blanket* of the *anterior thoracic wall* – whereas latero-caudally the *abdominal part* of this major chest-relief forming muscle is relatively weak (Fig. 8). Nevertheless, with its offspring from the anterior layer of the uppermost part of the rectus sheath (sheath fibers closely interwoven with costoxiphoid ligaments) this slip indicates the borders to both, the abdomen and the lateral thoracic wall (at the latter, lateral cutaneous branches of intercostal nerves emerge between the slips of the serratus anterior, which in turn intermingles with those of the external oblique; Fig. 8).

The most important structure in relation to the *anterior thoracic wall* is the *internal thoracic artery*, *ITA* (also – very unsuitably – named internal "mammary" artery). Springing off the inferior circumference of the subclavian artery opposite the thyrocervical trunk in most cases, it first runs downward, forward, and medially. When entering the thorax (i.e., the superior mediastinum; Fig. 13), the artery is crossed by the phrenic nerve. Contrary to the nerve, this largest of all thoracic wall arteries leaves the mediastinum by coursing deep to the intercartilaginous (internal intercostal) muscles about a finger breath lateral to the sternal border (regularly nearest to it in the second intercostal space; Figs. 9–11,

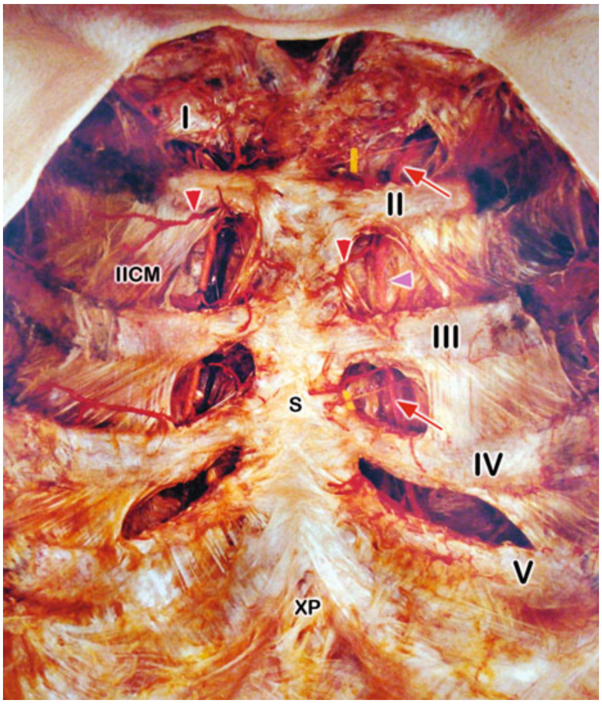

Fig. 9. Course of the internal thoracic artery (*ITA*) and vein (*ITV*) in "prepleural space" seen from anterior. The ITA (red arrows) is seen running parallel to the border of sternum (*S*) after partial removal of the internal intercostal muscles (*IICM*); terminal parts of the intercostal nerves (lined yellow) cross the ITA ventrally; the artery gives off perforating as well as sternal branches (read arrowheads); in the first two intercostal spaces, ITA and ITV (latter medial to artery) lie directly in front of the parietal pleura (pink arrowhead); I–V ribs; *XP* xiphiod process (covered by ligaments) (© Thiel W, Photographischer Atlas der Praktischen Anatomie II. Hals, Kopf, Rücken, Brust, Obere Extremität. Springer, Berlin Heidelberg, 1999, p 170, Fig. 85)

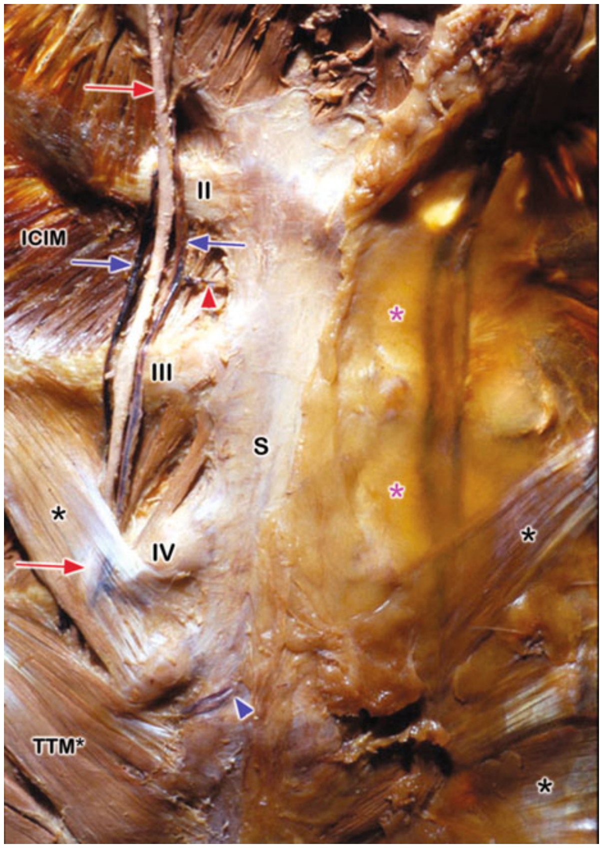

Fig. 10. Course of the internal thoracic artery (*ITA*) and veins (*ITVs*) in "prepleural space" seen from posterior. On the left side both parietal pleura and endothoracic fascia removed; in first two (to three, as seen here) intercostal spaces, ITA and ITVs (red and blue arrows) run directly in front of the parietal pleura (pink asterisks), whereas below they are covered by the transversus thoracis muscle (*TTM**); the vessels contact both, intercostales intimi muscles (*ICIM*) and costal cartilages (II–IV); note confluence of two ITVs to a single vessel is asymmetrical in this individual; red arrowhead: sternal branch of ITA; blue arrowhead: anastomoses between ITVs of either side on back of sternum (*S*).

14). The artery is usually crossed ventrally by the terminal parts of the intercostal nerves (before they pierce the muscles to become the anterior cutaneous branches).

Importantly, in the first two (sometimes three) intercostal spaces (ICS) the vessel is separated from the *parietal pleura* only by a very thin layer of connective tissue, the *endothoracic fascia* (Figs. 10 and 14), whereas below till the bifurcation at the sixth intercostal space (into superior epigastric and musculophrenic), the *transversus thoracis muscle* acts as a strong, muscular barrier! So for most of its intra-thoracic course the vessel lies outside the mediastinum in a space that could topographically be referred to as "*prepleural space*" (Figs. 10 and 14). Apart from the vessels, it contains parasternal lymph nodes and loose areolar tissue (Fig. 11); it is traversed by the transversus thoracis muscle. This space can be viewed in analogy to the retropleural space (Fig. 13), not least because of the endothoracic fascia: The endothoracic fascia is the only thing both, prepleural space and anterior mediastinum (see below), have in common, as it acts as the posterior cover of the former and the anterior of the latter (Fig. 14).

The *superior epigastric artery* is the continuation of the ITA which, very much in contrast to descriptions in most text books, does not (!) travel through the sternocostal triangle (also known as Larrey's space) but reaches the abdomen passing in front of a plane formed by both, the transversus thoracis and transversus abdominis muscles, to enter the posterior

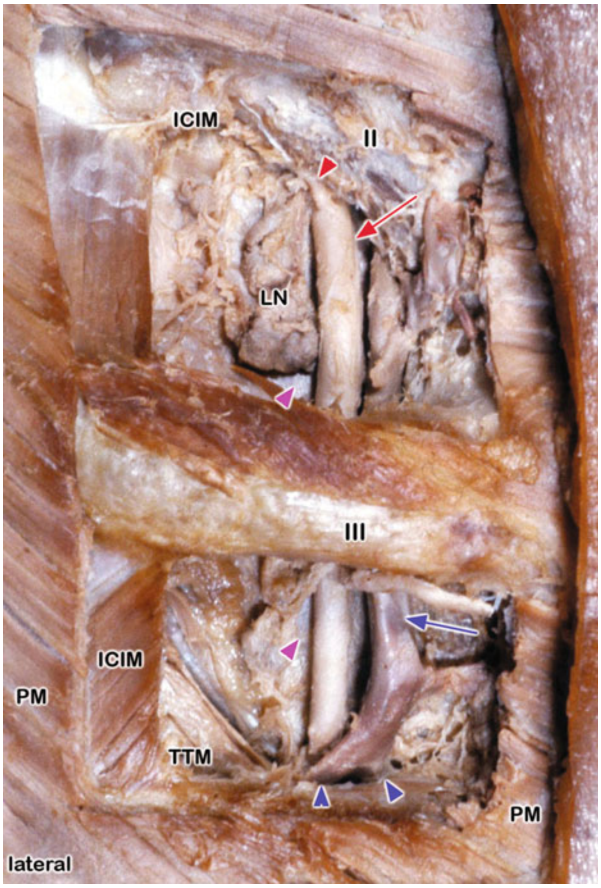

Fig. 11. Detail of internal thoracic vessels' topography with neighboring structures. Pectoralis major (*PM*) and intercostales intimi muscles (*ICIM*) fenestrated over second and third intercostal space; II and III costal cartilages; two venae comitantes (blue arrowheads) unite to from the internal thoracic vein (ITV, blue arrow) medial to the artery (red arrow); both lie on the parietal pleura (pink arrowheads) and endothoracic fascia (with loose areolar tissue) in the "prepleural space"; LN small, parasternal lymph nodes; red arrowhead: offspring of an anterior intercostal artery; TTM cranial border of transversus thoracis muscle

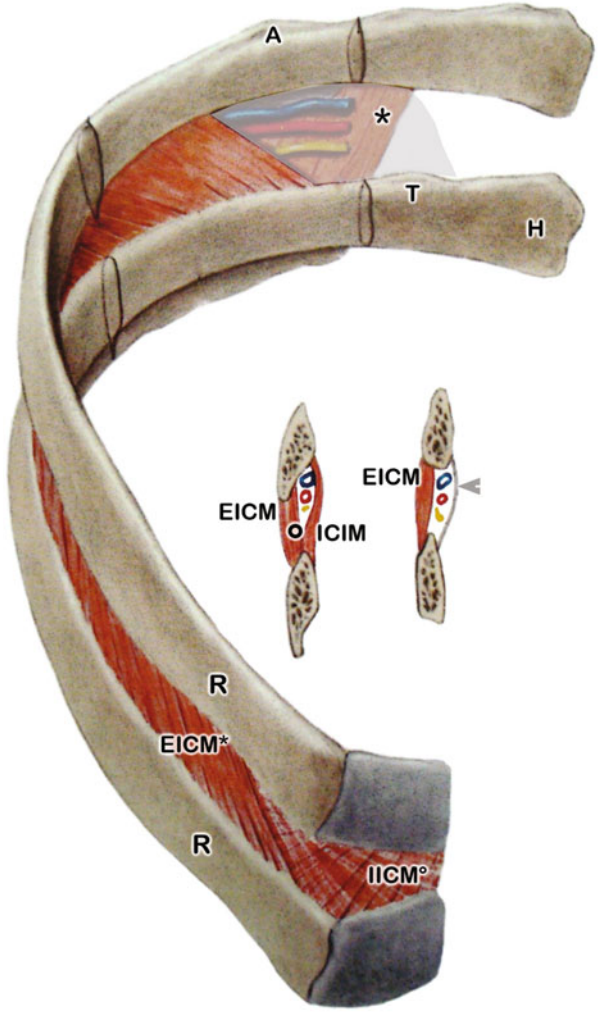

Fig. 12. Two ribs isolated with intercostal muscles and "Intercostal canal" with neurovascular bundle. *R* rib; *A* angle; *T* tubercle; *H* head; *EICM** external intercostal muscle; *IICM* internal intercostal muscle; *ICIM* intercostalis intimus muscle; insert cross section right: topographical situation medial to A; insert cross section left: antero-lateral to A; transparent grey and grey arrowhead: internal intercostal membrane (© Lanz T v, Wachsmuth W, Praktische Anatomie. Rücken. Springer, Berlin Heidelberg, Special Edition 2004, p 40, Fig. 55)

compartment of the rectus sheath. The *musculophrenic artery* (topographically more exact: costophrenic artery) runs in an oblique-downward direction between costal arch and respective origins of the diaphragm. That way it supplies the latter, abdominal muscles and the antero-lateral thoracic wall with anterior intercostal arteries to the seventh, eighth, and nineth intercostal spaces (two for each space – sometimes from a common stem – the cranial one usually of greater caliber).

In general, *branches* of the *ITA* supply the *anterior thoracic wall* and the *mediastinum* (superior and anterior), respectively.

The parietal branches are mainly represented by the *anterior intercostal arteries* for *ICS 1–6* (Fig. 11) which anastomose with the posterior intercostal arteries and their collateral intercostal branches (the latter run near the upper(!) margin of the lower rib of the upper six spaces). Similar to offsprings from the musculophrenic arteries, the two for each space may come from a single trunk, while sometimes such a stem will provide the two vessels above and below a single rib (that is for two adjacent ICSs). Uninflu-

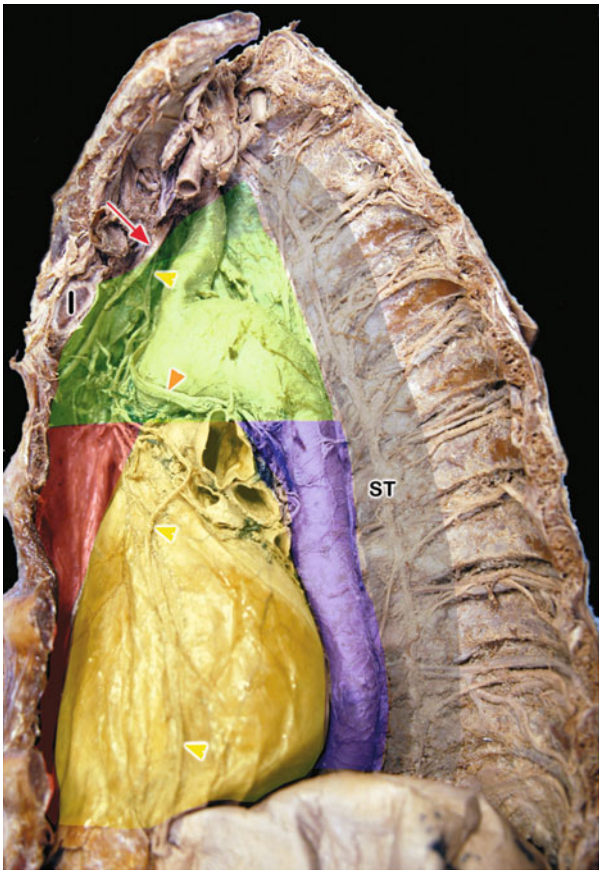

Fig. 13. Mediastinum and internal thoracic artery (*ITA*) relationships viewed from left (lung, pleura, and endothoracic fascia removed). The ITA (red arrow) enters the superior mediastinum (transparent green; other colored areas: inferior mediastinum, see below) crossed by the phrenic nerve (yellow arrowheads); after a short course the artery enters the "prepleural space" (no longer seen!) and lies outside(!) the anterior mediastinum (transparent red); in contrast, the phrenic nerve enters and runs within the middle mediastinum (transparent yellow) and the vagus nerve (orange arrowhead) reaches the posterior mediastinum (transparent purple); *I* first (left) rib; *P* pericardium; transparent grey: retropleural space with sympathetic trunk (ST)

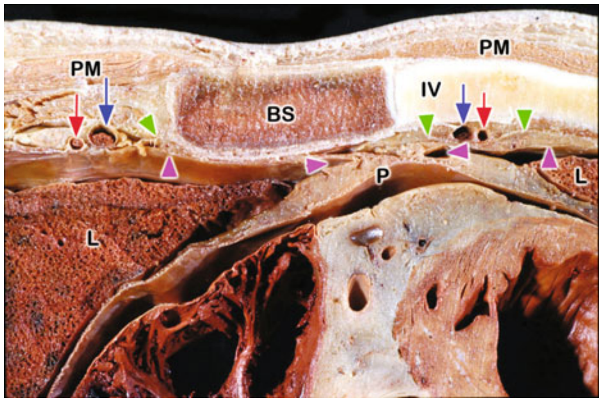

Fig. 14. Detail of cross section of anterior thoracic wall and anterior mediastinum (approximately mid-portion) at the level of seventh thoracic vertebra; seen from inferior. *PM* pectoralis major muscle; *BS* body of sternum; *IV* costal cartilage of fourth rib; *P* pericardium; pink arrowheads: parietal pleura and costomediastinal recesses of pleural cavity; note that right pleural sac almost reaches left sternal border! *L* lung; both internal thoracic vessels (red and blue arrows) in front of pleura ("prepleural space"); *note* only one vein on both sides! green arrowheads: endothoracic fascia

enced by variability of origin, they first course between parietal pleura and internal intercostal muscles and then run further laterally within internal intercostals to meet their "posterior counterparts". That way, arterial rings are formed in contact with the two borders of ribs bounding each of the ICSs. Apart from veins, the upper *arterial pathway* is accompanied by the main intercostal nerves, while their (smaller) collateral branches travel with the lower one. Due to greater dimensions, especially the upper of these *neurovascular bundles create* the two laminae of internal intercostals, the innermost known as intercostales intimi. Usually, the bundle in the upper compartment or "intercostal canal" is arranged from cranial to caudal in the order "V-A-N" (Fig. 12). The nearer to the sternum, the less developed is this topographically important *subdivision of the internal intercostal* muscles. Where present, this thin layer acts as the last barrier to the pleura outside the boundaries of the transversus thoracis muscle (see above!). Such typical separation is often completely absent in the first, sometimes also the second space. Additional parietal twigs of the ITA are the *sternal branches* (Figs. 9–10). These arteries connect to those from the contralateral side, especially on the posterior surface of the sternum(!), and are also nutrient vessels to the sternocostalis muscle. They either emerge individually from the ITA or from a common stem together with the *perforating branches* (Fig. 9) which finally reach the skin by piercing the pectoralis major muscle.

Among the visceral branches of the ITA, only the slender artery following the entire path of the phrenic nerve is named, the *pericardiacophrenic artery*. Because of its relationship with the nerve, it additionally supplies part of the pleura and is only close to the anterior thoracic wall at site of origin (posterior to the first intercostal space; Fig. 13). Other arteries of variable caliber reach the *thymus* (or its remains), the *pericardium* and *fat* adjacent to the anterior thoracic wall and the chain of *lymph nodes* accompanying the ITA and veins (Fig. 11).

Concerning the *internal thoracic* (mammary) *vein(s), ITV(s),* there is surprisingly little disagreement toward topography if various textbooks are taken as reference; the common (summarized) description being as follows: "The ITA is accompanied by two veins, *venae comitantes*, which unites at the third costal cartilage or second ICS to form a single vessel (ITV) lying medial to the artery".

It has to be emphasized that only the latter statement (vein medial to artery; Figs. 9–11) is tenable and logic, because the ITV ends in the corresponding brachiocephalic vein more medially compared to the ITA origin. In fact and summarized, there is (1) *remarkable variability* as to the assumed *level of confluence* of the two accompanying veins of each side to form the actual ITV: in more than 40% the confluence is clearly above or below the given description; (2) *notable asymmetry of* such *union* occurs in more than 50% of individuals (Fig. 10); and (3) *only one companion vein* throughout the course of the ITA on *one side* (meaning "overall asymmetry") is seen with a frequency of at least 10%, whereas a single ITV on both sides is rare.

Importantly (apart from many anastomoses between comitant veins of one side), there are numerous *anastomoses* across the back of the sternum to the contralateral side (Fig. 10)!

Addendum: A practically relevant *variant* of thoracic wall arteries *in males* is an aberrant lateral branch of the ITA that arises close to its origin: the *lateral internal thoracic artery*. Such an artery is not infrequently seen but often small and terminating at the level of ICS 2–3; however, it may (rarely) descend for the whole length of the *lateral thoracic wall* and may in caliber exceed that of the ITA proper!

Within the context of *thoracic wall vessels* in general, both anterior and lateral, it should finally be underlined that most of their practically relevant anatomy – including variability as mentioned – may easily be *evaluated by* means of *Color Duplex Sonography*.

2.1.4.2 Thoracic cavity: the (anterior) mediastinum (see Figs. 13 and 14)

The major portion of the thoracic cavity is occupied by both lungs in their *pleural sacs*. Per definition, the *mediastinum* is what remains *in between* the medial surfaces of the parietal pleura (mediastinal part). This central portion of the cavity has two major components, superior and inferior; the latter *subdivided* by the *pericardial sac* in posterior, middle (heart as main contents), and anterior (Fig. 13).

The anterior mediastinum reaches from the posterior surface of the *body of* the *sternum* in front to the anterior surface of the *pericardium behind* and is laterally bounded by the *costomediastinal recesses* of the pleura (Fig. 14). That means, because of the most variable extension of the pleural sacs with often close approximation (or even overlapping) between the level of second and fourth costal cartilages, it may be exceedingly narrow (or absent) in its upper part and only representing a true space in the lower; there, the lines of junctions of mediastinal and costal pleura diverge to form the "cardiac triangle". Moreover, the anterior lines of pleural reflection often considerably shift away from the midline so that the right pleural sac may reach (or exceed) the left sternal border (Fig. 14!), and vice versa! Even in those extreme cases, the internal thoracic vessels will still run outside the anterior mediastinum ventral to the costal pleura: "prepleural space" (see above)!

For the most part, the anterior mediastinum is filled with *loose areolar, fatty tissue*. Other structures are: *the sternopericardial ligaments* (membranous and very variable as to their ligamentous character or "strength") a maximum of two to three *small lymph nodes* and small *branches of* the *ITA* (and veins). Parts of the *thymus* (remains) may or may not lie within the upper portion of the anterior mediastinum. This is mainly depending on the described behavior of the pleura on the one hand and *age* on the other! After puberty, the organ will progressively diminish in size and thus draw itself more and more back into the superior mediastinum, whereas in childhood it covers a great area of the mid-portion of the ventral pericardium, thus being an essential part of the anterior mediastinum.

2.2 Genetics of chest wall deformities

Dieter Kotzot

Chest wall deformities are observed as a single anomaly or as a symptom of various monogenic syndromes, numeric and structural chromosome aberrations, genetic associations, and disruption sequences (Tables 1–3). For chest wall deformities as a single anomaly etiology and/or pathogenesis are not known and from clinical experience the recurrence risk will be low in most cases. In contrast to some syndromic entities for isolated anomalies no molecular studies to elucidate the genetic background have been published up to now. For most instances subtypes relevant for surgical procedures were not even discussed separately in medical genetics. In the following the most frequent and therefore most important monogenic syndromes (Marfan syndrome and Noonan syndrome), disruption sequences (Poland anomaly and Moebius anomaly), genetic associations (Pentalogy of Cantrell and PHACE), and isolated chest wall deformities such as pectus excavatum and carinatum and cleft sternum will be reviewed in more detail with respect to physicians and specialists dealing with treatment and diagnosis of these deformities.

2.2.1 Pectus excavatum and carinatum

Pectus excavatum or funnel chest (= depression of the sternum/adjacent ribs) and pectus carinatum (= protrusion of the sternum/adjacent ribs) have been described as a single anomaly or as a symptom of various monogenic syndromes, numeric or structural chromosome aberrations, or disruption sequences. Pectus excavatum is the most frequent anterior chest wall deformity. Incidence is around 1:400 in live births with a male-to-female ratio of ca. 4:1 [5]. The embryological basis for these anomalies is not clear. Abnormal growth of the costal cartilages is considered to be causative, however, so far triggers for and pathogenesis of abnormal growth are not known [21]. Familial non-syndromic (= isolated) pectus excavatum (OMIM 169300) has been reported rarely and mainly in the former literature [14, 19, 23]. As in these reports clinical documentation is often poor and inheritance is considered to be autosomal dominant, mild Marfan or any other monogenic syndrome cannot be excluded. Recently, the first but however only descriptive family study was published. Creswick et al. [6] analyzed 34 families and assumed autosomal dominant inheritance in 14 families, autosomal recessive inheritance in 4 families, and X-chromosomal inheritance in 6 families. However, the number of cases was too small for statistical evaluation, and moreover, the authors noted that many family members had additional connective tissue traits. So, in most of these families a systemic connective tissue disease cannot be excluded. A search in POSSUM (*P*ictures Of *S*tandard *S*yndromes and *U*ndiagnosed *M*alformations version 5.7) and WBDD (*W*inter-*B*araitser *D*ysmorphology *D*atabase version 1.0.14), two widely used expert systems for the diagnosis of rare genetic syndromes, resulted in 39 monogenic syndromes, 8 numeric chromosome aberrations, and 44 structural chromosome rearrangements (Tables 1 and 2). The first group includes metabolic disorders, connective tissue disorders, skeletal dysplasias, and classical dysmorphic syndromes. The most important and most frequently observed monogenic syndromes associated with pectus excavatum and carinatum are Noonan syndrome (OMIM #163950) and Marfan syndrome (OMIM #154700), respectively.

Pectus excavatum (Fig. 1) or carinatum is a hallmark of Marfan syndrome, a systemic disorder of connective tissue, which is caused by mutations in the *Fibrillin 1* gene localized on the long arm of chromosome 15 (reviewed by Le Parc JM, Orphanet 2005). Prevalence of this autosomal dominantly inherited disorder is around 1:5–10,000. Penetrance is almost 100% with broad inter- and intrafamiliar variability ranging from isolated features to severe presentation already in neonates, and poor genotype-phenotype correlation. Recurrence risk for a patient's children is 50%, but almost 25% of all mutations are de novo. A clinical diagnosis of Marfan syndrome is possible, if aortic root aneurysm and ectopia lentis are present, or in the absence of either of these two, the presence of

Table 1: Proven or assumed monogenic syndromes with pectus carinatum and/or pectus excavatum listed in POSSUM (version 5.7) or WBDD (version 1.0.14) (the most frequent disorders with regard to frequency or clinical relevance are in bold)

Syndrome	Inheritance	OMIM	Location	Gene
Acropectorovertebral (F-) syndrome	AD	102510	2p36	
Al-Gazali et al. [1] – webbed neck–facial dysmorphism–congenital heart disease	AR			
Becker nevus syndrome	?	604919		
Brachycephaly – deafness – cataracts – mental retardation	?	601353		
Camptodactyly – type Guadalajara	AR	211910		
Carbohydrate deficient glycoproteins syndrome IIA	AR	212065	14q21	CDGS2
Cardiofaciocutaneous (CFC) syndrome	**AD**	**115150**	**12p12.17q34**	**KRAS, BRAF, MEK1, MEK2**
Catel-Manzke – cleft palate; accessory metacarpal	?	302380		
Coffin-Lowry syndrome	X-linked	303600	Xp22	RSK2
Contractural arachnodactyly (Beals)	AD	121050	5q23–5q31	FBN2
Desbuquois syndrome	AR	251450	17q25.3	
Dundar et al. [8] – acropectoral syndrome	AD	605967	7q36	
Facio-auriculo-thoracic syndrome	AR			
Hirschsprung disease – microcephaly–mental retardation	AD		2q22	SIP1
Holt-Oram syndrome	**AD**	**142900**	**12q21-2414q23-24?**	**TBX5**
Homocystinuria	AR	236200	21q22	CBS
Hypomelanosis of Ito	X-linked (?)	300337		
King-Denborough – dysmorphic features; myopathy; malignant hyperthermia	AD	145600	19q13.1	RYR1
Kyphomelic dysplasia	AR	211350		
Lehman et al. [13] – osteosclerosis; abnormalities of nervous system/ meninges	AD	166720		
LEOPARD syndrome	AD	151100	6q12q24	PTPN11, RAF1
Loeys et al. [15] – aortic aneurysm, hypertelorism, arterial tortuosity, CP	AD	190182609192	3p229q33–q34	TGFBR1, TGFBR2
Lowry-Wood syndrome (epiphyseal dysplasia, microcephaly, nystagmus)		226960		
Marden-Walker syndrome	AR	248700		
Marfan syndrome	**AD**	**154700**	**15q21**	**FBN1**
Marfan syndrome – severe neonatal	AD	154700	15q21	FBN1
Mucopolysaccharidosis type VII	AR	253220	7q21.11	MPS7
Multiple epiphyseal dysplasia with Robin phenotype		601560		

(*Continued*)

Table 1: (*Continued*)

Syndrome	Inheritance	OMIM	Location	Gene
Multiple epiphyseal dysplasia–microcephaly–nystagmus	AR	226960		
Mutchinick syndrome		249630		
Nevoid basal cell carcinoma syndrome (Gorlin)	AD	109400	9q22-q31	PTCH1
Noonan syndrome	**AD**	**163950**	**12q24.112p12.2p223p25**	**PTPN11, KRAS, SOS1, RAF1**
Noonan-like/multiple giant cell lesion syndrome	AD	163955	12q24.1	PTPN11
Occipital horn syndrome	X-linked	304150	Xq13	MNK
Osteogenesis imperfecta types I, III, IV	**AD, AR**	**166200259420166220**	**17q22, 7q22.1**	**COL1A1, COL1A2**
Pseudoachondroplasia	AD	177170	19p13.1	COMP
Shprintzen-Goldberg – arachnodactyly; craniosynostosis, hernias	?	182212	3p2215q21	TGFBR2, FBN1
Somlo et al. [22] – marfanoid syndrome with polycystic renal disease	AD		16p13	
Three M syndrome	AR	273750	6p21.1	CUL7

AD autosomal dominant; *AR* autosomal recessive

Table 2: Chromosome aberrations associated with either pectus carinatum or pectus excavatum listed in POSSUM (version 5.7) and WBDD (version 1.0.14) (the most frequent aberrations with regard to frequency or clinical relevance are in bold)

Del 1q41->42	Diaphragmatic hernia, lung hypoplasia, microcephaly, coarse face with (mostly) full lips, bulbous nasal tip, prominent forehead, and deep-set eyes
Dup 1q	Pre- and postnatal growth retardation, cardiac defects, mental retardation, macrocephaly or micro-/brachycephaly, facial dysmorphisms (broad/high forehead, depressed nasal bridge, and downslanting palpebral fissures), wrinkled skin, Pierre-Robin sequence, pulmonary hypoplasia, and flexion contractures
Mosaic trisomy 1	CNS, cardiac and lung defects, overlapping and flexed fingers, facial dysmorphisms (hypertelorism and prominent eyes)
Del 2p	Mental and postnatal growth retardation, prominent broad nasal bridge and bulbous nose, high-arched palate, micrognathia, and anomalies of fingers/toes
Del 2q13->q21	Developmental delay, microcephaly, corpus callosum defects, cardiac anomalies, prominent forehead, low-set and malformed ears, tendency to recurrent severe infections
Del 2q37	Moderate–severe mental retardation, short stature, rounded face with short nose and flattened nasal bridge, short metacarpals and metatarsals
Dup 2p	Short stature, microcephaly, severe mental retardation, aortic stenosis, high prominent forehead, hypertelorism, depressed nasal bridge, long/narrow trunk and scoliosis, soft skin, and hyperextensible fingers with arachnodactyly
Dup 2q	Growth and mental retardation, cardiac, renal and GI anomalies, facial dysmorphisms (hypertelorism, depressed nasal bridge, short nose, long philtrum, low-set ears, and micrognathia)
Del 3p25->pter	Severe mental retardation, congenital heart disease, kidney defects, placid personality, pre-/postnatal growth retardation, asymmetric skull and face, telecanthus, ptosis, micrognathia, low hair line, synophrys

(*Continued*)

Table 2: (*Continued*)

Del 3p12->p14	Profound growth and mental retardation, cardiac abnormalities, annular pancreas, renal anomalies, and facial dysmorphisms (hypertelorism, ptosis, epicanthus (inversus), broad and high forehead, broad flattened nasal bridge)
Del 3q29	Moderate mental retardation, growth delay, hypotonia, horseshoe kidney, hypospadias, and facial dysmorphism
Mosaic trisomy 4	Cutis marmorata, frontal bossing, hirsutism, and short neck
Del 5p15	**Severe growth and mental retardation, hyperextensible joints, triangular face, and multilobulated ear tags**
Del 5qter	Mild developmental delay, macrocephaly, bell shaped chest, brachydactyly, dysmorphic facies with telecanthus and anteverted nares
Del 6p25	Cardiac defects, developmental delay, anterior chamber anomalies, hypertelorism, downward slanting palpebral fissures, smooth philtrum, and deafness
Del 6p22->24	Brain, heart and kidney abnormalities, short neck, facial dysmorphism, clinodactyly, or syndactyly
Del 6q	Micro-/brachycephaly, absent pulmonary valve, flat face, hypertelorism, bulbous nose, and malformed ears
Del 6q27	Developmental delay, autism, seizures, hypotonia, mild microcephaly, enlarged ventricles, absent corpus callosum, ear anomalies (prominent, protruding, and large), epicanthal folds, and flat philtrum
Dup 6p	Severe mental retardation, low birth weight, microcephaly, hypertelorism, ptosis, prominent nasal bridge, small mouth, pointed chin, and low-set ears
Del 7p21->pter	Craniosynostosis, urogenital and cardiac defects, facial dysmorphisms, mental retardation, anomalies of hands and feet
Del 7p15.3->21.2	Craniosynostosis, mental and growth retardation, and craniofacial abnormalities
Del 7q11.21	**Supravalvular aortic and/or peripheral pulmonary artery stenoses, elfin-like hypotonic face with thick lower lip, large mouth, long smooth philtrum, periorbital fullness, stellate iris pattern, full cheeks (jowls), and dental anomalies, constipation and feeding difficulties, hypercalcemia in infancy, moderate growth and developmental delay, outgoing personality**
Recomb 8	Mental retardation, cardiac and genito-urinary abnormalities, dysmorphic facies
Del 8pter	Congenital heart malformations, microcephaly, IUGR, mental retardation, and characteristic hyper-active impulsive behavior
Del 8p	Pre-/postnatal growth retardation, microcephaly, narrow forehead, epicanthus, moderate–severe mental retardation and congenital heart defects
Dup 8q	Severe growth and mental retardation, congenital heart disease (particularly conotruncal anomalies), absent gall bladder, renal anomalies, skeletal anomalies, and facial dysmorphism (hypertelorism, broad forehead, triangular face, downslanting palpebral fissures, broad nasal bridge, and long philtrum)
Mosaic trisomy 8	**Cardiac, renal and skeletal defects, large babies with deep palmar and plantar creases, coarse expressionless face with thick lips, prominent ears, absent/dysplastic patellae, velopharyngeal insufficiency, and mild to moderate mental retardation (normal intelligence has been reported)**
Del 9q22.1->q22.32	Dysmorphic and mentally handicapped
Del 9q22,3	High birth weight and macrocephaly (also trigonocephaly), mental retardation, triangular face, frontal bossing, epicanthal folds, small mouths, and thin upper lips
Del 9q34	Hypotonia, developmental delay, natal teeth, single umbilical artery, microcephaly, facial dysmorphism (epicanthic folds, downslanting palpebral fissures, hypoplastic midface, and small nose with depressed nasal bridge)
Tetrasomy 9p	IUGR, ventriculomegaly, contractures, renal anomalies, mental retardation, hypertelorism, beaked/bulbous nose, and cleft lip/palate
Del 10q26	Mental disability, growth retardation (pre- and/or postnatal), ano/genital defects, cardiac- and renal anomalies, microcephaly, triangular face, hypertelorism, strabismus, prominent nasal bridge, low-set ears, micrognathia, and short neck
	(*Continued*)

Table 2: (*Continued*)

Dup 10q	Mental and growth retardation, skeletal, heart and renal anomalies, microcephaly, round and flat face, high-arched eyebrows, downslanting short palpebral fissures, and tented upper lip
Del 11q	Heart and urogenital defects, thrombocytopenia, trigonocephaly, hypertelorism/telecanthus, short nose, microretrognathia, carp shaped mouth
Del 12q	Pyloric stenosis, growth and developmental delay, hypertelorism, ptosis, low-set and rotated ears, and sparse hair
Del 12q24	Moderate mental retardation, bouts of aggressive behavior, normal growth, microcephaly, prominent forehead, hypoplastic supraorbital ridges, long eyelashes, deep-set eyes, strabismus, paranasal broadening, unilateral cleft lip and palate, large mouth with cupid-bow shaped upper lip, and small ears
Dup 12p	Ulnar deviation, developmental delay, cardiac anomalies, shawl scrotum, facial dysmorphisms (hypertelorism, flat nasal bridge, micrognathia, low-set rotated ears)
Trisomy 13	Brain malformations (holoprosencephaly), cleft lip/palate, polydactyly, and variable organ defects
Mosaic trisomy 14	Growth and mental retardation, congenital heart disease, micropenis/undescended testes, body asymmetry, streaky/linear hyperpigmentation, facial dysmorphisms (hypertelorism, wide nasal bridge, micrognathia, cleft/high-arched palate, low-set/dysplastic ears), and short neck
Del 15q15->q22.1	Craniosynostosis, facial dysmorphism, severe mental retardation, tetralogy of Fallot, limb anomalies, facial dysmorphisms (hypertelorism, beak-like nose, hypoplastic alae nasi, thin upper lip, micrognathia), late-onset obesity, scalp defect
Dup 15q	Postnatal growth and mental retardation, bulbous nose, arachno-/camptodactyly, cardiac and genital defects
Del 17p13	Severe mental retardation, postnatal growth deficiency, hypotonia, seizures, microcephaly, cortical atrophy, partial agenesis of the corpus callosum, facial anomalies, long fingers, and bilateral talipes equinovarus
Dup 17p	Pre-/postnatal growth, mental retardation, heart defect, microcephaly, hypertelorism, downslanting palpebral fissures, thin upper lip, and micrognathia
Dup 17q	Growth and mental retardation, microcephaly, frontal bossing, widow's peak, downturned mouth, short neck, hirsutism with sparse scalp hair
Del 18p	Growth and mental retardation, seizures, skeletal and genital defects, facial dysmorphisms (hypertelorism, epicanthic folds, wide mouth with downturned corners, and single maxillary incisor)
Del 18q12.1->q21.1	Mental retardation, abnormal behavior, obesity, dysmorphic features (high/prominent forehead, deep-set eyes, hypotelorism, short midface, short nose, and flat philtrum)
Dup 18p11	Pre- and postnatal growth deficiency, developmental delay, microcephaly, round face, hypertelorism, small nose, low-set and dysplastic ears, preauricular pits, sensorineural hearing loss, small chin, swallowing difficulties, VSD, and III/IV cutaneous syndactyly
Trisomy 21	**Cardiac and intestinal defects, peculiar facial grimacing with tongue thrusting, hypotonia and delayed motor milestones, increased risk for leukemia**
Monosomy X	**Include congenital lymphedema of hands and/or feet, short stature, short/webbed neck, low posterior hairline, cubitus valgus, nail hypoplasia, broad chest, infertility, left-sided cardiac anomalies**
Dup Xq	Multiple congenital anomalies including mental retardation, short stature and facial dysmorphisms (short palpebral fissures, ptosis and downturned mouth)
49,XXXXY	Mental retardation, cleft palate or bifid uvula, coarse facies, radioulnar synostosis, hypogenitalism and cardiovascular defects, verbal skills extremely poor
Tetraploidy	Small baby with joint contractures, dysmorphic facies, and multiple congenital anomalies

Del deletion; *dup* duplication

a bonafide Fibrillin1 mutation or a combination of systemic manifestations (Fig. 2) is required [16]. The involvement of various organ systems requires a multidisciplinary approach in diagnosis and therapy. In up to 91% of patients meeting these criteria a mutation in the *Fibrillin 1* gene can be detected by molecular methods [17]. A related phenotype of arachnodactyly, aortic root aneurysms, pectus deformi-

Table 3: Chromosome aberrations monogenic syndromes, and clinical associations associated with cleft sterum listed in POSSUM (version 5.7) and/or WBDD (version 1.0.14) (the most frequent disorders with regard to frequency or clinical relevance are in bold)

Syndrome	Inheritance	OMIM	Location
Asternia			
Cantrell's Pentalogy	X-linked, sporadic	313850	Xq25-26.1
Bohring et al. [4] midline body wall defects – facial anomalies			
C syndrome	AR	211750	3q13
Chromosome 9q22.3 – submicroscopic deletion			9q22
Dandy-Walker malformation – facial hemangiomas	AR	220200	
Ectopia cordis – cleft lip/palate			
Goltz (focal dermal hypoplasia)	**X-linked**	**30560**	**Xp22, 9q32-34**
Miles-Carpenter – X-linked MR; fingertip arches; contractures	X-linked	309605	Xq21
Ozlem et al. [20] – anophthalmia – anal atresia – rhizomelia	AR		
Perlman syndrome (gigantism with renal dysplasia/tumors)	AR	267000	
Pterygium colli medianum – midline cervical cleft			
Pulmonary agenesis-unilateral	AR	601612	
Split hand/foot-tibial defects	AR, AD	119100	
Sternal clefts – telangiectasia/hemangiomas		**140850**	
Teruel et al. [25] – absent abdominal musculature, microphthalmia, joint laxity	AR		
Uygur et al. [26] – omphalocele, ectopia cordis, absent tibia, oligodactyly			
Van Allen-Myhre – ectopia cordis; split hand/foot; skin defects	AR		
Yunis-Varon – cleidocranial dysostosis plus	AR	216340	
Amniotic band syndrome		217100	
Coffin-Lowry syndrome	**X-linked**	**303600**	
Congenital hypothyreoidism	**AR, AD**	**218700**	
PHACE syndrome		**606519**	

AD autosomal dominant; *AR* autosomal recessive

ties, scoliosis, dural ectasia, and facial dysmorphisms but no eye involvement (Loeys-Dietz syndrome) is caused by mutations in the transforming growth factor-beta receptor type I or II (*TGFBR1* and *TGFBR2*) (OMIM 190182) [15].

Noonan syndrome is a common autosomal dominantly inherited disorder caused by various genes in the RasMAPK (*M*itogen *A*ctivated *P*rotein *K*inase) pathway (reviewed by Allanson [2]). The RAS (*RA*t *S*arcoma viral oncogene homolog) proteins and their downstream pathways are a signaling cascade important for cell proliferation, differentiation, survival, and cell death. *PTPN11* (*P*rotein *T*yrosine *P*hosphatase *N*on-receptor type 11) is mutated in ca. 50%, *SOS1* (son of seveless gene 1) in ca. 10%, *RAF1* (v-raf1 murine leukemia viral oncogene homolog 1) in 3–17%, and *KRAS* (Kirsten RAS) in ca. 5% of patients, but further genes are assumed to be responsible for Noonan syndrome. Most mutations are hypermorphic resulting in an increased and prolonged signal flux. Genotype-phenotype correlation is only weak. Incidence is 1:1,000–2,500. The phenotype is characterized by normal measurements at birth, short stature later in life, congenital heart defects (totally 50–80% most frequently pulmonary valve stenosis (20–30%)) and/or cardiomyopathy (20–30%), broad or webbed neck, cranial pectus carinatum and caudal pectus excavatum inferiorly (Fig. 4), cryptorchidism (60–80%), coagulation defects, and facial dysmorphisms including ptosis, wide-spaced eyes, and low-set and posteriorly rotated ears [2]. Some of these features are due to jugular lymphatic obstructions. Mental development is variable. Learning disabilities have been reported in 25% and further 10–15% need special education. Treatment of medical problems is symptomatic and the same as in the general population. Differential diagnosis includes Turner syndrome, Williams-Beuren syndrome (OMIM 194050), Aarskog syndrome (OMIM 305400), Cardio-

Fig. 1. A 13-year-old girl with Marfan syndrome and asymmetrical pectus excavatum deformity, predominately at the left side. She also exhibits cardiopulmonal impairment due to the severe sunken sternum deformity

Fig. 2. Same patient as in Fig. 1 with typical arachnodactylia also featuring a Marfan syndrome

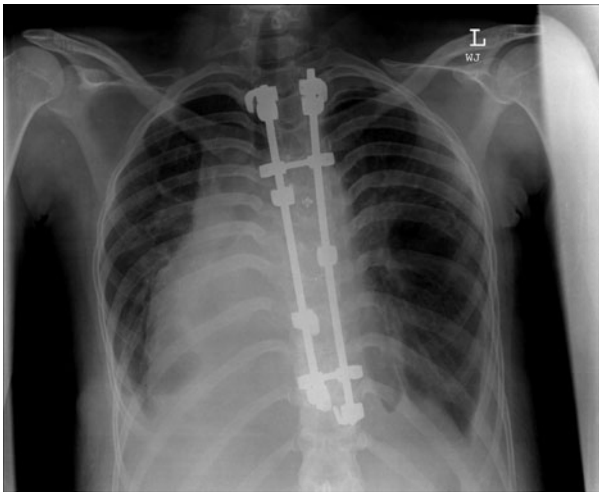

Fig. 3. Female patient with Noonan syndrome and dextrocardia. The former scoliosis of the vertebral column is repositioned by internal stabilization. She also underwent closure of open ductus Botalli and reconstruction of congenital abdominal wall dysplasia

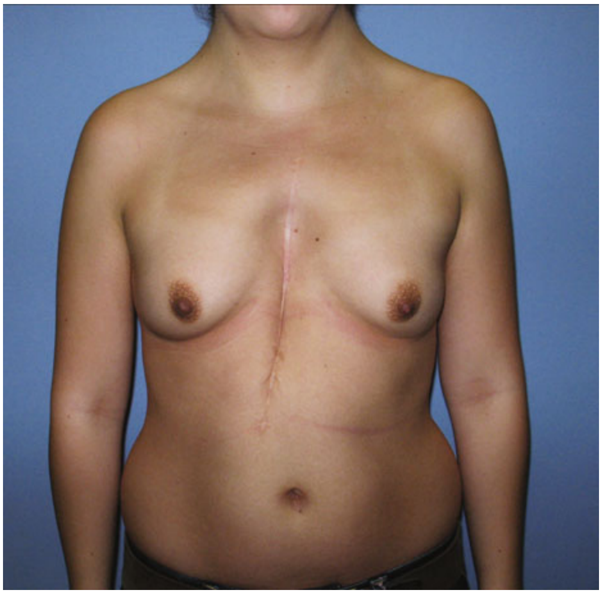

Fig. 4. Another female patient with Noonan syndrome, noticeably a webbed neck, a cranial pectus carinatum with transition into an excavatum deformity caudally. The presternal scar results from atrial septum and pulmonal valvular surgery in early childhood

faciocutaneous syndrome (OMIM 115150), Costello syndrome (OMIM 218040), and LEOPARD syndrome (multiple *L*entigines, *E*lectrocardiographic conduction abnormalities, *O*cular hypertelorism, *P*ulmonary stenosis, *A*bnormalities of genitalia, *R*etardation of growth, and sensoneural *D*eafness) (OMIM 151100). For a proportion of the last three syndromes also mutations in the RasMAPK pathway have been described. Due to the autosomal dominant inheritance recurrence risk for children of affected patients is 50%.

Furthermore, there are reports of few or even single families with multiple congenital anomalies and pectus excavatum as an additional obligatory feature. Zori et al. [28] described a family with pectus excavatum, macrocephaly, short stature, dysplastic nails and an apparently autosomal dominant inheritance (OMIM % 600399). Beals and Fraser [3] reported on a 3-generation family with bowing of the tibia, pseudarthrosis, and pectus excavatum (OMIM %609143). Khaldi et al. [12] reported on two brothers born to consanguineous parents with short stature, mental retardation, pectus excavatum, and camptodactyly. Guízar-Vázquez et al. [10] described a brother and his sister with a peculiar face, pectus carinatum, and joint laxity. The latter both entities are hints toward an autosomal recessive inheritance.

Poland anomaly (OMIM 173800) and Moebius anomaly (OMIM 157900) are considered as disruption sequences. Most cases are sporadic. Chest wall deformities in Poland anomaly mainly include Sprengel anomaly as well as hypoplastic and fused ribs (Fig. 5). Additional features include hypoplasia or absence of nipple or even the entire breast, hypoplasia or absence of the pectoralis major muscle, hemivertebrae, and brachy-, syn-, and oligodactyly. All features are unilateral and more often affecting the right than the left side. Incidence is around 1:32,000 and male-to-female ratio is 2:1 to 3:1 [9]. Various theories of etiology have been reported (disruption of the lateral plate mesoderm, subclavian artery supply disruption, etc.). Bilateral symptoms and an almost equal male-to-female ratio have been observed in few familial cases reported so far. The latter indicates an autosomal dominant inheritance in these families and thus a recurrence risk of 50% in offsprings. For unilaterally affected patients the recurrence risk for children is low. Poland anomaly can be associated with Moebius anomaly, which is characterized by unilateral or bilateral congenital facial and abducens palsies and additional features like micrognathia, epicanthic folds, dysplastic ears, defective branchial musculature, and various limb defects [27]. Due to lingual involvement, general motor disabilities, poor coordination, and respiratory abnormalities Verzijl et al. redefined Moebius anomaly as a syndrome of rhombencephalic maldevelopment. In rare families autosomal dominant, autosomal recessive and X-linked inheritance have been assumed (reviewed by Verzijl et al. [27]), but most cases are sporadic and recurrence risk is low. Cleft sternum can be observed as a single anomaly or as a symptom of various monogenic syndromes and chromosome aberrations. It is a rare malformation caused by a failure of ventral cell migration and fusion of sternal bands in the sixth to ninth week of gestation [21]. Variability ranges from an indentation in the manubrium to a complete non-fusion of sternal bands. It is frequently associated with a supraumbilical raphe resembling a postoperative scar and keloid. Particularly in women it had been noted with cavernous hemangiomata. A search in POSSUM (version 5.7) and WBDD (version 1.0.14) resulted in 22 monogenic syndromes, and 1 structural chromosome rearrangement (Table 3). Most cases of non-syndromic cleft sternum are sporadic, but familial cases have been reported, so in general the recurrence risk is low. Haque [11] described two affected siblings born to consanguineous parents. The boy showed complete cleft sternum, while his sister had a superior cleft sternum and a left-sided facial cavernous hemangioma. Clefts of the lower part of the sternum are found in the Pentalogy of Cantrell (OMIM 313850), which, in addition, is characterized by a midline supraumbilical abdominal wall defect resulting in an omphalocele, diastasis of the recti, or even an absent umbilicus, deficiencies of the diaphragmatic pericardium and the anterior diaphragm, and a cardiac abnormality like an ASD, a VSD, an ectopic heart, a double outlet right ventricle, a truncus arteriosus, or an anomalous pulmonary venous drainage [24]. Most cases are sporadic with a low recurrence risk, but occasionally

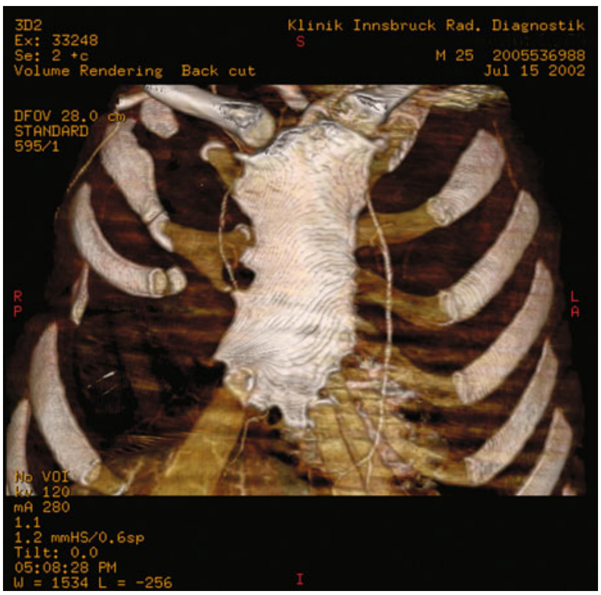

Fig. 5. Male patient with Poland anomaly and partially absent and fused anomalous ribs at the right side as well as a pectus carinatum deformity at the same side, caused by rib cartilage protrusion and slight malrotation of the sternum

X-chromosomal inheritance with a gene on Xq25-26.1 was assumed. PHACE (*P*osterior fossa brain malformations, *H*emangiomata of the face (large or complex), *A*rterial anomalies, *C*ardiac anomalies, and *E*ye abnormalities) (OMIM 606519) is a neurocutaneous association [18]. Ventral development defects, particularly sternal defects and/or supraumbilical raphe has been reported. Etiology and pathogenesis are unknown. In a study of 1,096 children with infantile hemangiomata 25 patients met the criteria for PHACE. Most cases are sporadic indicating a low recurrence risk, and nearly 90% of all patients are female.

2.2.2 Summary

Chest wall defects can be an isolated malformation or dysmorphic feature or only one symptom of a genetic syndrome. From clinical experience the recurrence risk of non-syndromic chest wall deformities is low. Investigations of the genetic and biochemical basis of isolated pectus excavatum, pectus carinatum, and cleft sternum are still at the beginning, but new technical methods like high-throughput sequencing or whole genome association studies with high-density SNP-arrays are promising tools for the next future. At the moment, every patient with a chest wall deformity should be carefully evaluated for additional symptoms not only of the skeleton but also of other organ systems. If any genetic syndrome is suspected, the patient should be referred for genetic counseling to confirm the syndromic diagnosis and to discuss possible molecular investigation, recurrence risk and clinical variability in offsprings, and related issues. An interdisciplinary approach is also recommended to choose the best therapeutic approach for each patient.

Glossary

Deformation	morphological anomaly as a consequence of a mechanical force
Disruption	morphological anomaly as a consequence of an external agent
OMIM	Online Mendelian Inheritance in Man (http://www.ncbi.nlm.nih.gov/omim/)
Penetrance	proportion of patients with a clinical phenotype out of a group of mutation carriers
Syndrome	pattern of anomalies or malformations as a direct consequence of one common cause

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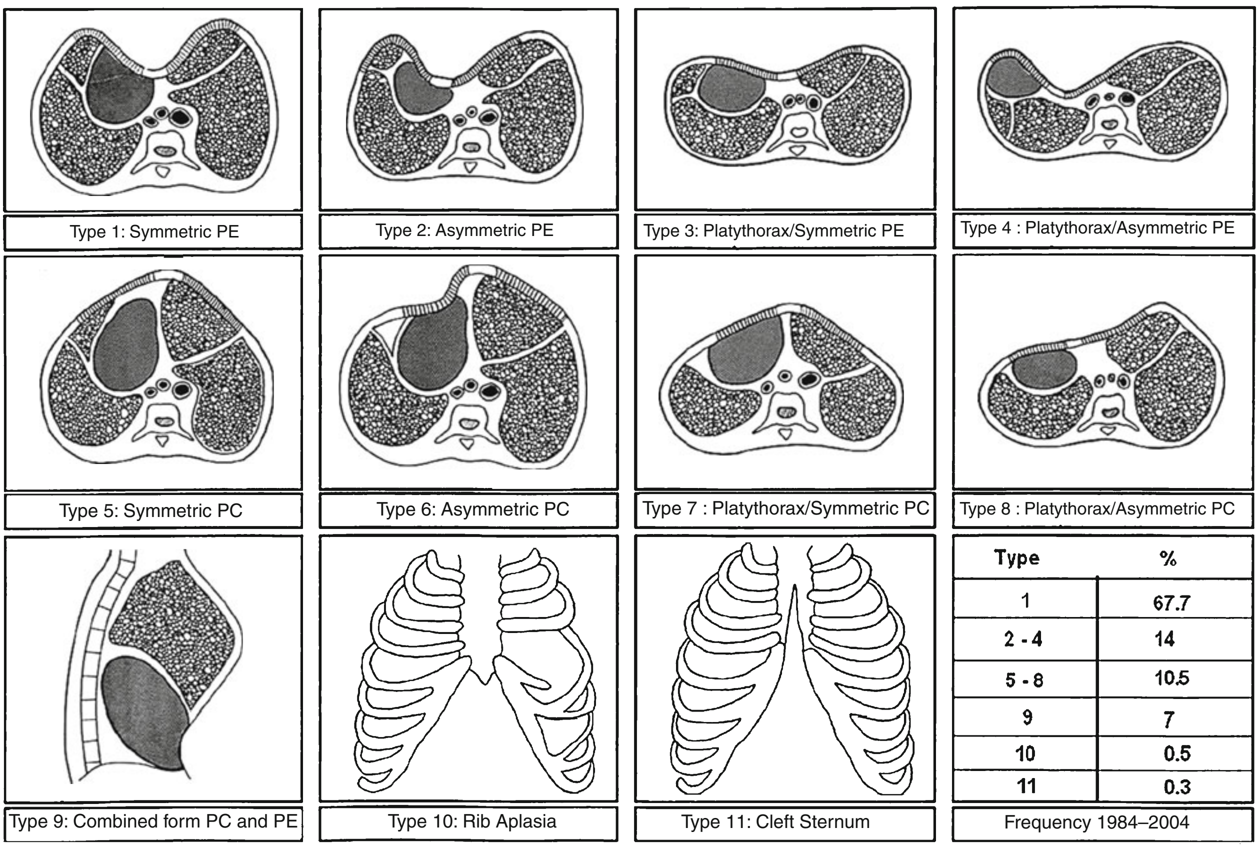
POSSUM: Murdoch Children’s Research Institute, Royal Children’s Hospital, Parkville, Victoria, AustraliaWBDD: London Medical Databases Ltd, London, UK

2.3 Classification/definition/description of typical and rare deformities

Barbara Del Frari, Anton H. Schwabegger

Congenital or developmental deformities of the anterior chest wall do not primarily result in severe functional problems as in major limb anomalies. The appearance is also much better tolerated and more easily camouflaged exemplarily compared with major facial clefts or other congenital malformations. That is why apart from highly specialized physicians who place particular emphasis on dealing with such deformities, minor or little attention has been paid so far throughout general medicine until the advent of the MIRPE technique. Since then a myriad of publications was edited and the deformity attracted significant interest with newly constructed options of diagnosis, classifications, and treatment. Apart from the factor of surgeons's experience, the patient's factor, which means a diversity of pectus morphology, influences the rate of complications and success of outcome [8, 10]. It is therefore of paramount importance to be informed in detail about which different expressions of deformities exist and how their morphology will guide the selection of an appropriate treatment method [11, 12]. Fundamentally a classification presents itself then as simple when the differences between the entities are



Type	%
1	67.7
2 - 4	14
5 - 8	10.5
9	7
10	0.5
11	0.3

Frequency 1984–2004

Fig. 1. Scheme depicts a clear and distinctive classification of the anterior chest wall deformities (reprinted from: The Journal of Cardiovascular Surgery, Vol. 134, No. 4, by Saxena AK, Willital GH; "Valuable lessons from two decades of pectus repair with the Willital–Hegemann procedure"; pp. 871–876, 2007, with permission of Elsevier)

distinct thus can be clearly circumscribed. If there exist many transitions between the extreme forms of pectus excavatum and those of pectus carinatum, the classification yet turns out to become somehow sophisticated. Above all the separation and integration of a variety of mixed forms appear to become challenging when the classification process remains a matter of subjective estimation. The classification proposed by Willital (Fig. 1) on the other hand appears to be meaningfully pragmatic for clinical use in the respect that it is restricted to few types of excavatum and carinatum deformities likewise, incorporates asymmetric versions, and is relatively easily surveyable based on the distinctive features described [15, 17]. Several other classifications have been made so far, and usually are based on the anatomical variations in terms of the relation of the dislodged sternum and distorted rib cartilages out from a physiological frontal plane and considering the possible asymmetry [2, 3, 9, 10, 14]. These classifications however are limited to a segment of the whole variety of anomalies of the anterior chest wall, either circumscribing the excavatum or the carinatum deformity. From a holistic view with respect to treatment options, it seems

Table 1: The Willital classification

Type 1	Symmetric pectus excavatum within a normal configured thorax
Type 2	Asymmetric pectus excavatum within a normal configured thorax
Type 3	Symmetric pectus excavatum associated with platythorax
Type 4	Asymmetric pectus excavatum associated with platythorax
Type 5	Symmetric pectus carinatum within a normal configured thorax
Type 6	Asymmetric pectus carinatum within a normal configured thorax
Type 7	Symmetric pectus carinatum associated with platythorax
Type 8	Asymmetric pectus carinatum associated with platythorax
Type 9	Combination of pectus excavatum and pectus carinatum
Type 10	Thoracic wall aplasia
Type 11	Cleft sternum

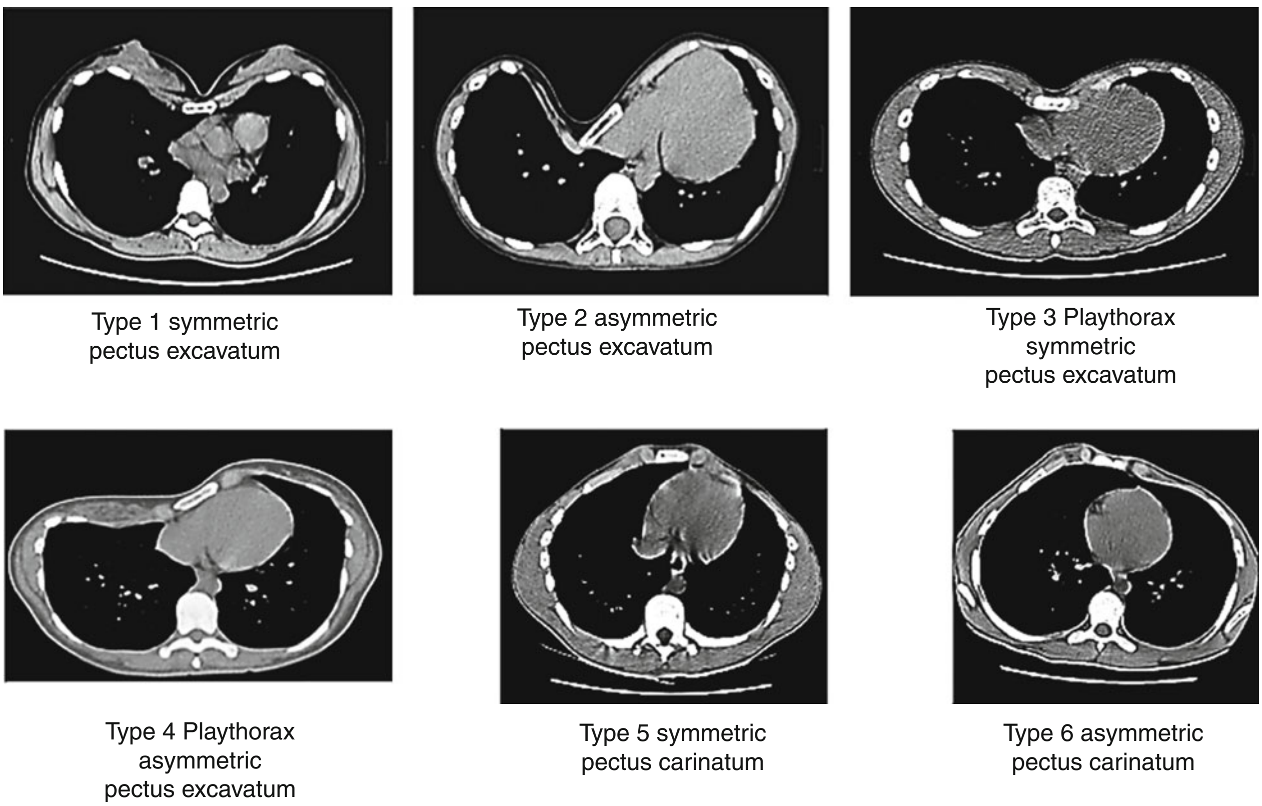


Fig. 2. CT scans from a variety of anterior chest wall deformities also showing the extent of sternum malrotation predominantly present in the asymmetric cases

more practicable to simplify and reduce [15, 17] any such classification to an utmost necessary number of deformities (Table 1) instead of incorporating detailed minor form variants that might confuse the patient as well as the physician. One issue however that significantly influences the selection of adequate treatment is the absence or presence of sternum malrotation, which may be expressed to a very variable extent in almost every type of classified deformity thus must be described in addition with indexing a present deformity. Such malrotation (synonymous twisting or torsion) of the sternum out of the frontal plane above 30° is estimated as severe [3] thus must be considered during planning of the surgical procedure. Major sternum distortions may complicate the minimally invasive procedures as well as the open access likewise by rigid resistance against remodeling procedures (Fig. 2).

The type "pectus arcuatum" is the corresponding deformity No. 9 according to Willital and is described there as a combined form of pectus excavatum and carinatum along a longitudinal axis. However, as pointed out by Robicsek in 1979 [13] one should distinguish between the pouter pigeon breast and the asymptomatic pectus arcuatum deformity (Chapter 2.3.3). This is of importance as the pouter pigeon breast, consisting of a protrusion based on premature ossification [4] at the angle of Louis in about 30% is associated with organic heart defects [5, 13].

An exception of these entities represents the Poland syndrome, which will be described in detail in Chapter 2.3.4. In the Poland syndrome, predominantly absence of muscles and soft tissue at the chest wall as well as varieties of underdevelopment at the upper extremity is a matter of concern and treatment (Chapter 10). In several cases of Poland syndrome, the chest wall also shows a variety of deformities such as absent ribs, hemithoracal hypoplasia, or asymmetrically developed keel chest. In such cases, the description of Poland syndrome should be appropriately complemented with a circumscribed specification of the chest wall deformity rather than merging with any classification types.

Another rare entity described by Spear in 2004 is an anterior thoracic hypoplasia, consisting of a sunken thoracic wall unilaterally, hypoplastic female breast, superiorly placed nipple areola complex but normally developed pectoralis muscles [16]. Mühlbauer and Wangerin in 1977 [8] named a Poland syndrome which is associated with unilateral female breast aplasia or severe hypoplasia the Amazone syndrome (Chapter 2.3.4). Just by citing here two rare entities, one may argue and many physicians certainly already encountered rare transitional forms of these descriptions, which cannot be classified into a common scheme. As such entities with more or less not definitely classifiable characteristics [1] occur very rarely apart from well-defined types of deformities, and they should be summarized as mixed deformities.

The variety of the deformities of the anterior chest wall seems to have different anatomical [3] and biomechanical causes. Its understanding should implicate further studies on the genetics and biomechanical development in order to adapt future treatment to the etiology [7] thus potentially minor invasive access. Delayed ossification of the sternum with several present partitions (Fig. 3 in Chapter 2.3.6) may contribute to the development of pectus deformities. In a radiologic study Haje in 1999 stated that endochondral growth of sternum as well as of the cartilaginous rib arches influences the development of pectus carinatum and asymmetric pectus excavatum deformities, based on the anatomic concept that sternum and costal growth is based upon growth plates and disturbances therein may develop during body maturation [6]. However, no correlation in sternal development could be found with extensive pectus excavatum deformities. The findings in this radiologic study cited that enchondral ossification and growth plates to a variable extent contribute to the type of deformity and thus should be considered in the future treatment of pectus deformities. Suggested epiphysiodesis instead of chondrectomies at the affected rib cartilages and sternal growth plates might therefore be a considerable modern access for minor invasive treatment [6].

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2.3.1 Funnel chest/pectus excavatum and subgroups

Barbara Del Frari, Anton H. Schwabegger

The pectus excavatum deformity (in Latin: pectus = breast; ex = out of; cavare = to hollow out), also known as funnel chest, is characterized by a depression usually involving the lower one-half to two-thirds of the sternum. There exist many variants concerning the extent of such a deformity, from minimally thus hardly recognizable forms to extents with severe dislodging of intrathoracic organs by narrowing the sterno-vertebral distance to almost zero. The depression inclinates at the manubriogladiolar junction (angle of Louis) and usually reaches the deepest point at the xiphisternal junction (Figs. 1–4). The superior portion of the manubrium, the first and second rib pairs, and the corresponding cartilages are usually spared. Although the affected cartilages are inwardly curved, the ribs lateral to the costochondral junctions often remain unaffected. There may be an additional component of sternal twisting, also named sternum malrotation (Figs. 5–8). In up to 50% of excavatum deformities, the sternum is twisted out of a frontal plane and is turned to the right more frequently than to the left [3, 5].

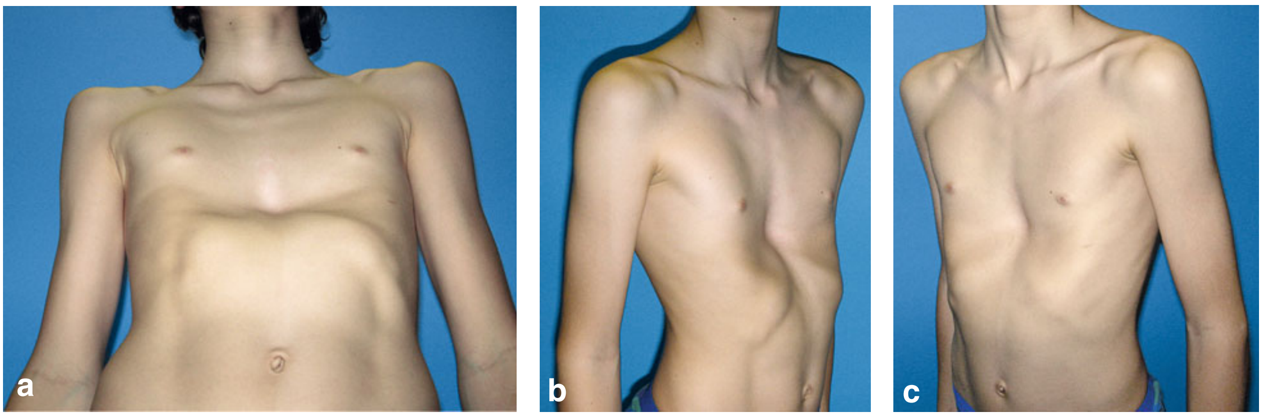

Fig. 1a–c. Almost symmetrically shaped pectus excavatum

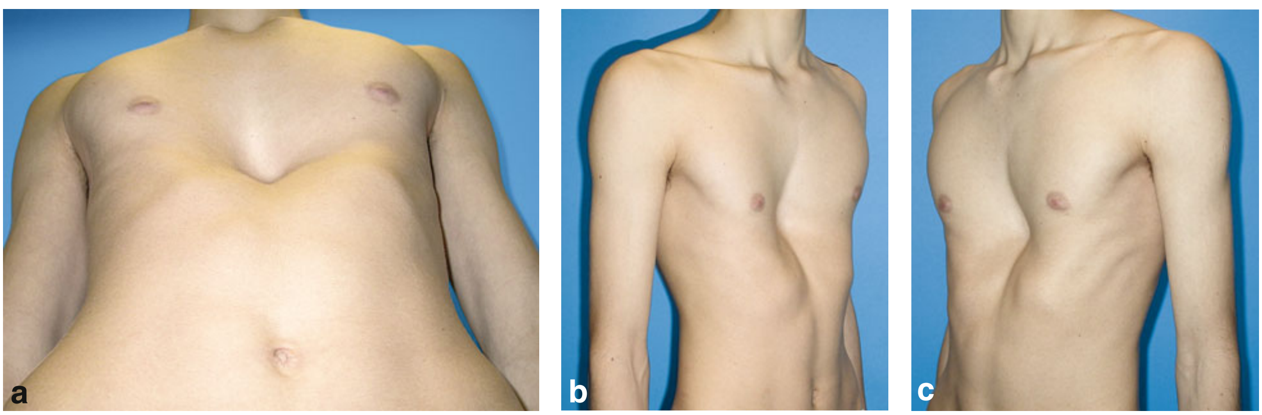

Fig. 2a–c. Symmetrically shaped pectus excavatum, but with deep depression of sternum

Fig. 3. Almost symmetrically shaped pectus excavatum, with asymmetry at the lower rib arches

Fig. 4. VR three-dimensional CT imaging with perfect symmetry of the thoracic cage in severe pectus excavatum

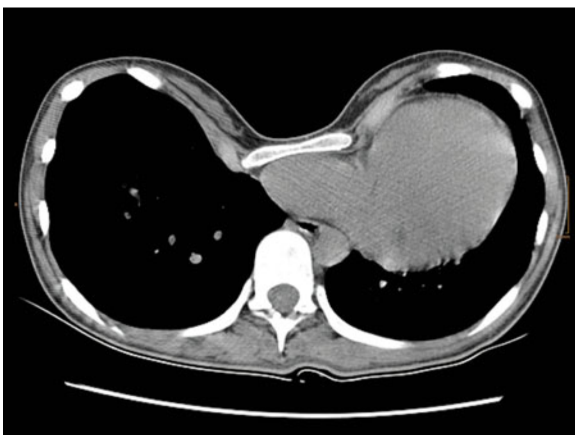

Fig. 5. Conventional CT scan with moderately asymmetric pectus excavatum and indicated minor platythorax

Fig. 6. VR three-dimensional CT imaging with severe asymmetric pectus excavatum and platythorax

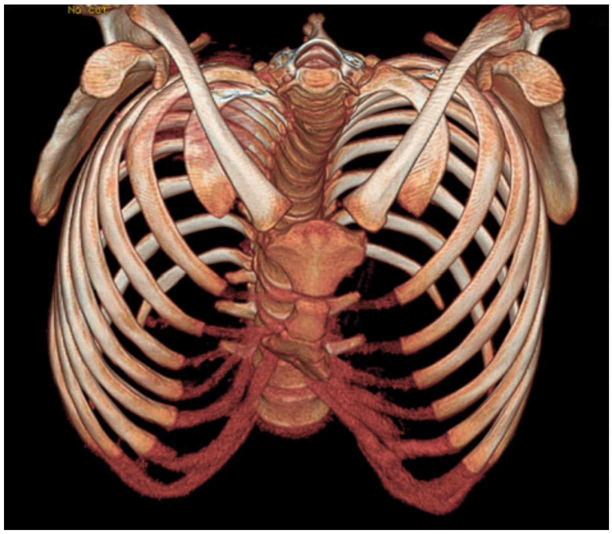

Fig. 7. VR three-dimensional CT imaging with asymmetric pectus excavatum and sternum malrotation

A classification system within the pectus excavatum deformities has been proposed by several authors [1, 4, 7, 11, 12]. The primary division is between symmetric and asymmetric deformities. Symmetric deformities are characterized by co-localization of the center of the depression with the median line of the sternum (Figs. 1–4). Asymmetric deformities (Figs. 9–13) are differently described in the medical literature based on the type of concomitant thoracic deformity as described exemplarily by Saxena and Willital [10, 12]. A combination of different types of deformities or only minor expressions with exemplarily anterior thoracic hypo-

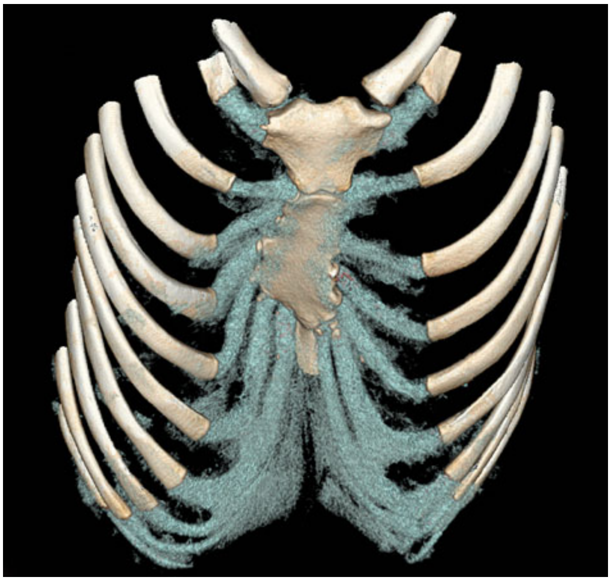

Fig. 8. VR three-dimensional CT imaging with massive asymmetric pectus excavatum in a female with Marfan syndrome

Fig. 9. Asymmetric pectus excavatum with minor depression at the left side

plasia (Fig. 14) leads to a variety in the shape of a deformed anterior thoracic wall, which can hardly fit into a rigid grid of classification.

Many of other multiplanar deformities, as long as they do not clearly meet the requirements of an understandable description, should be summarized as mixed deformities (Chapter 2.3.7) including an additive description to its dominant feature.

Pectus excavatum is the most common congenital chest deformity. It has a reported incidence of between 1:1000 and 8:1000 live births [10], with a male-to-female ratio ranging from 3:1 [10] to 9:1 [6].

Fig. 10. Asymmetric pectus excavatum with minimal depression at the right side but sternum malrotation

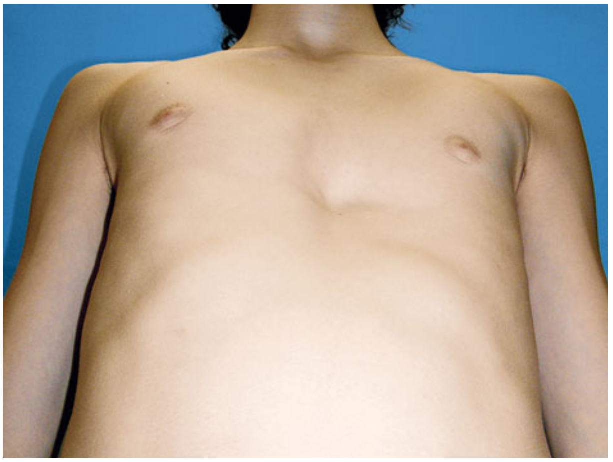

Fig. 11. Asymmetric pectus excavatum without depression at the right side but sternum malrotation

Although majority of patients with pectus excavatum are recognized during the first year of life, the depression usually increases during the period of rapid skeletal growth first in childhood and then ensued in early adolescence apparently during growth spurts. Kelly in 2008 in a detailed survey pointed out the time of fusion of ossificated sternal cartilage parts. The so-called lower sternebrae normally fuse at the prebubertal age, whereas the upper three sternebrae fuse between puberty and the 25th year [5]. This knowledge implicates on the time of surgery but furthermore on the type of surgery. Therefore elevation of the sternum with unbending of its pathological curvature with respect to these findings of ossification patterns may be feasible

Fig. 12. Asymmetric pectus excavatum and platythorax, with minor sternum malrotation. Notice that the seeming breast asymmetry is caused by the chest wall asymmetry and not by asymmetry of breast volume

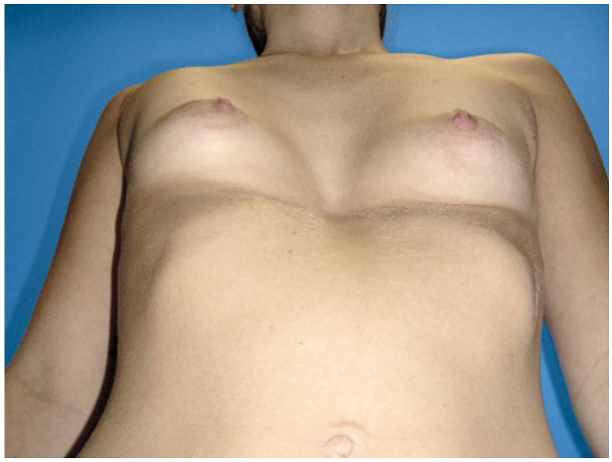

Fig. 13. Deeply shaped asymmetric pectus excavatum with sternum malrotation and protruding lower rib arches. Breast malposition at the patient's right side due to underlying severe skeletal asymmetry falsifies breast volume asymmetry

with internal support (i.e. metal struts) only until the age of 25 years. After the moment of complete ossification, only sternotomies should enable unbending attempts. When deformity progresses beyond the adolescent years, it is characteristically associated with lumbodorsal scoliosis in up to 65%, slumped shoulders and a protruding abdomen [3] usually caused by an inappropriate attitude to camouflage the "nasty" deformity.

Pectus excavatum deformity is characterized by various degrees of severity [8]. It frequently presents not only as an aesthetic disturbance but also in association with mild limitation of activity, slight to marked dyspnea, obstructive pulmonary mechanics, asthma, abnormal cardiac physiology, and palpitations [5, 6, 8]. In up to 95% of cases however, the deformity itself presents exclusively as an aesthetic burden, at times with associated deformities, syndromes, or connective tissue

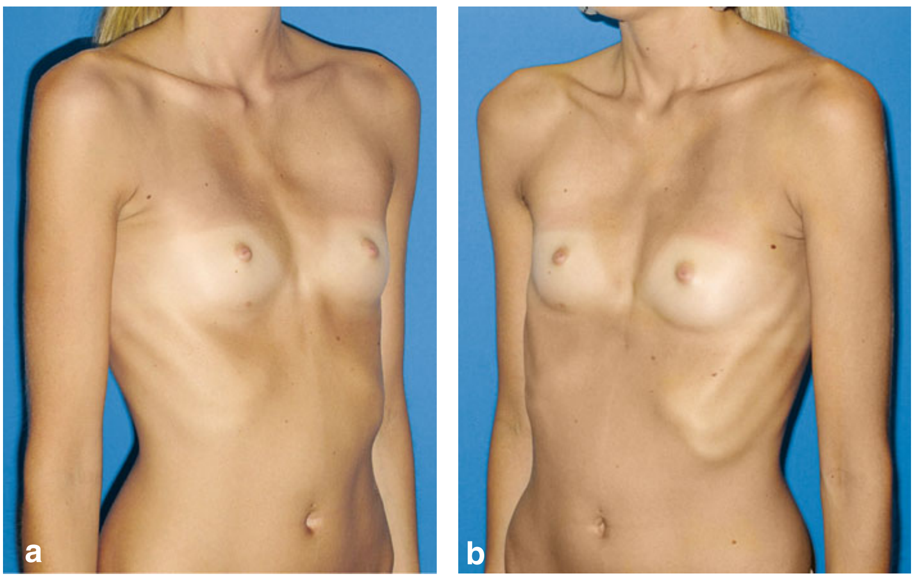

Fig. 14a and b. Symmetric deformity with minor excavatum features at the upper chest wall, not fitting into any classification grid, named anterior thoracic hypoplasia and consisting of platythorax and female breast hypoplasia disorders [5] such as a "hunched over" condition, dorsal kyphosis, thoracic scoliosis (15–21%), Marfan syndrome (5–8%), Ehler-Danlos syndrome (3%), or Sprengel deformity (0.6%).

Ravitch already decades ago [8] reviewed recorded experience with pectus excavatum. The reported cardiac disabilities include decreased return of blood to the right heart, cardiac arrhythmias secondary to atrial compression, restriction of diastolic filling, and a decrease of respiratory reserve. These findings however are limited to patients with severe pectus excavatum deformity with reduction of the sagittal sterno-vertebral distance.

Nevertheless, such cases with definite and concomitant cardiopulmonary symptoms requiring treatment remain a minority. Furthermore this incidence is not limited to childhood and may also occur in late adulthood, when skeletal rigidity increases by nature thus the flexibility to adapt respiratory volume diminishes and pulmonary hypofunction becomes apparent. Chest wall depression is well tolerated in infancy and symptoms seem to deteriorate until a steady state of completed pubertal growth has been achieved. Most pectus deformities maintain the same degree of severity throughout adult life, but based on increasing skeletal rigidity will gradually become more symptomatic [2]. Except for severe chest wall deformities, it is generally extremely difficult to predict the course of progression after a deformity has been recognized in early childhood, yet it may be a cumbersome ethic endeavor to suggest surgical correction during childhood, when no apparent cardiopulmonal malfunction is yet detectable, neither may be predicted with certainity for any higher age.

Apart from rare indications to widen the intrathoracic space for restoration of cardiopulmonal function, a classification of the predominantly aesthetic and psychically disturbing deformities seems to implicate the choice of surgical technique [5, 9]. That is, a classification allows assessment of the operative technique exemplarily with regard to the location of eventually necessary implantation of metal struts, as well as the optional determination of the number of struts to be used for internal stabilization of the chest wall [5, 11]. It also takes into consideration the asymmetric presentation of pectus deformities, along with inclusion of the rare forms. Evaluation of a study [11] with inclusion of 1262 patients with pectus deformities demonstrated that 67.7% of the patients have pectus excavatum deformity with a symmetric chest wall. Nevertheless the more rare asymmetric forms represent the surgically challenging collective thus the knowledge of such a distorsion of the sternum is of importance for a proper planning with adequate surgery.

An extremely valuable tool for classification as well as for therapy planning, but apart from prior accurate clinical examination (Figs. 1–14), is imaging of the deformity utilizing a three dimensional VR (volume rendering) CT-scan with particular respect to cartilages and sternum (Figs. 4–8).

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2.3.2 Keel chest/pectus carinatum and subgroups

Barbara Del Frari, Anton H. Schwabegger

Pectus carinatum (in Latin, "chest with a keel"), or pigeon breast, is the term applied to various protrusion deformities of the anterior chest. The disorder encompasses a spectrum of protrusion anomalies with much greater variation than that observed with the excavatum deformities.

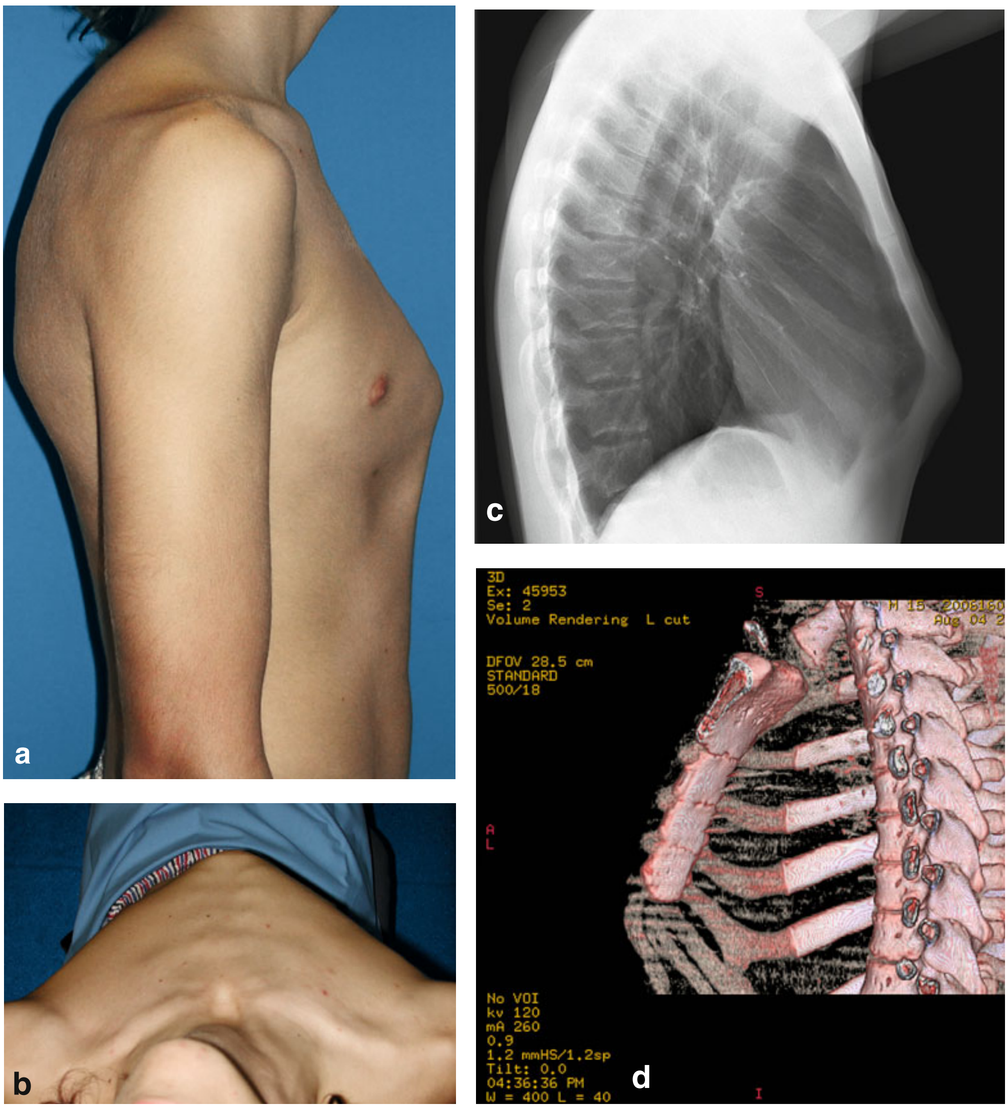

Fig. 1a–d. Extensive chondrogladiolar keel chest deformity in a 15-year-old male with protruding sternum at the lower part (**a**), almost symmetrically shaped (**b**). Lateral X-ray depicts the disfigured shape and extent of protrusion of the sternum (**c**). VR-CT scan of another patient also shows disfigured rib cartilages conjoined to the disfigured sternum (**d**).

The diversity of appellations concerning the pectus carinatum deformity ranges from keel chest, pigeon breast, pyramidal chest, sternum cuneiforme, and several others [7]. Such a variety of descriptions prove the existence of many expressions and different shapes, many of them not fitting into an attempted classification up-to-date [8, 11]. Nevertheless, an accurate description and diagnosis is mandatory as the three-dimensional architecture demands for different surgical approach and performance of remodeling of the affected part of the thoracic wall.

Brodkin in 1949 recognized and described two separate types of protruding chest deformities, the chondrogladiolar and the chondromanubrial shape of the anterior thoracic wall [1]. The aetiology still remains unknown and is ascribed to overabundant growth of the length of the rib cartilages. The most common, the chondrogladiolar type comprises anterior displacement of the mid and lower sternum (gladiolus sterni) and its attached costal cartilages (Fig. 1a–d), often asymmetric and usually first recognized during early adolescence. Asymmetric deformities are also

Fig. 2. Major asymmetric pectus carinatum deformity with twisted sternum (malrotation)

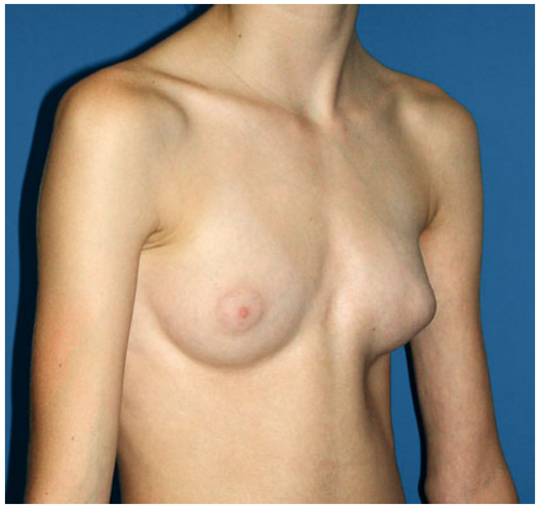

Fig. 4. Another female of 15 years of age with a similar but lesser extent of the deformity as in Fig. 3

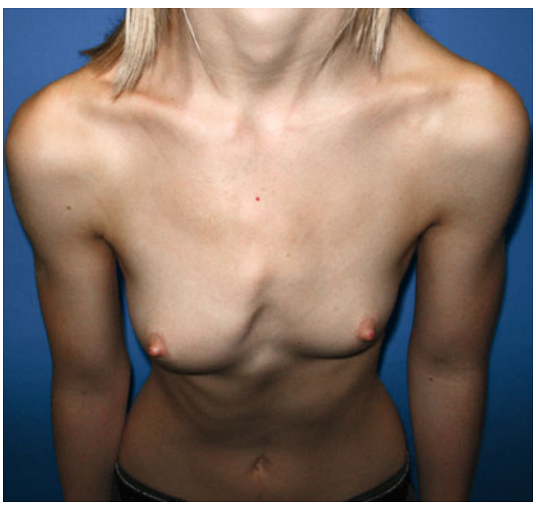

Fig. 3. Asymmetrically shaped pectus carinatum in a 17-year-old female at the middle third of the sternum with malrotation and lower thoracic wall depression at the patient's left side

Fig. 5. 49-Year-old male with minimal form of pectus carinatum but lower thoracic depression areas

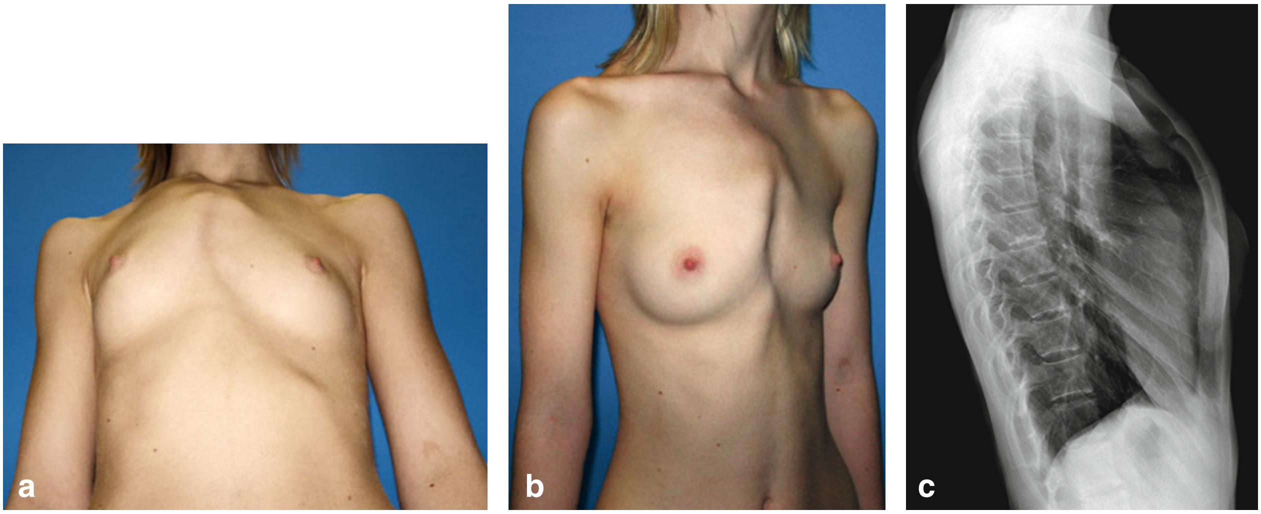

Fig. 6a–c. 16-Year-old female with asymmetric chondromanubrial deformity predominantly extending the middle third of the sternum and adjacent ribs (**a**, **b**). The X-ray depicts the location of the maximal sternal protrusion (**c**)

Fig. 7. 26-Year-old female with chondromanubrial deformity, hardly accessible for surgical correction from a concealed area to hide any scar

common and often involve a tilted sternum (Figs. 2 and 3), with localized depression of a few cartilages of the lower anterior lateral chest on one side (Fig. 4) or occasionally both sides (Fig. 5). Least common is a chondromanubrial (Figs. 6a–c, 7 and 8) (manubrium sterni) prominence, with protrusion of the manubrium, the upper sternum, and adjacent cartilages but less protrusion of the lower parts. Commonly this kind of deformity appears during the first few years of life. A chondromanubrial deformity with an occasional depression of the lower sternum occurs in approximately 15% of patients [2]. The latter deformity with an arcuate sternum is named pectus arcuatum and is described in Chapter 2.3.3, as well as a premature ossification of the sternomanubrial transition zone, which is called the pouter pigeon breast. In all types, the cartilages adjacent to the sternum, particularly rib cartilages two through six are malformed on both sides.

Robicsek et al. [6] noted a further type of pectus carinatum, the lateral pectus carinatum, either expressed unilaterally or bilaterally (Figs. 9 and 10a, b). This deformity is rather rare.

Keszler in 1999 noticed that out of 420 operations 21% were pure protrusions (pectus carinatum) and 15% were mixed deformities. In 4% only the mixed deformity was limited to the parasternal region, whereas in 13% the deformities extended to the entire anterior thoracic wall [4]. Up to 26% [10] of relatives of patients with pectus carinatum deformities show familial predilection. The overall incidence varies within several studies but is

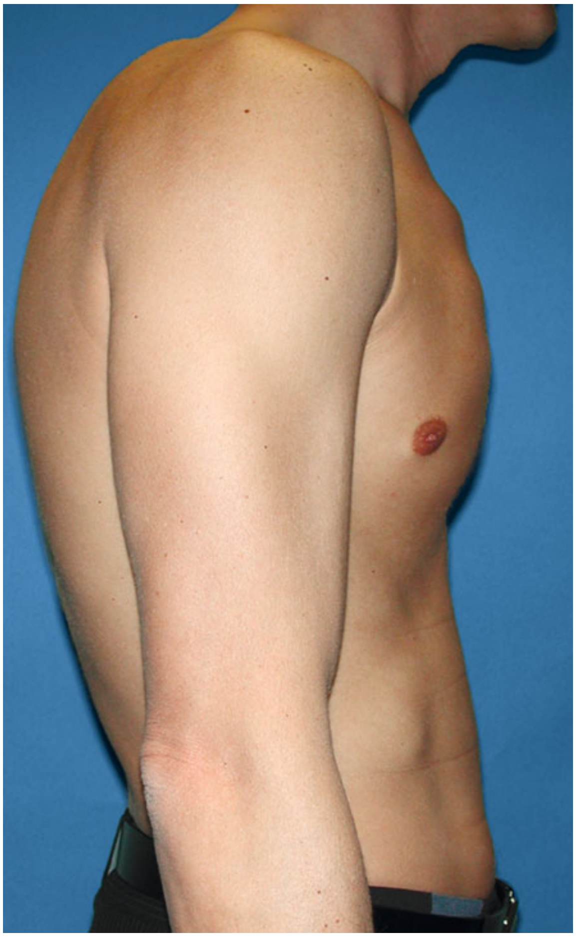

Fig. 8. 34-Year-old male with extended protrusion at the chondromanubrial region, extending down to the middle third of the sternum

Fig. 9. Unilaterally shaped pectus carinatum deformity with minor malrotation of the sternum especially at its lower part

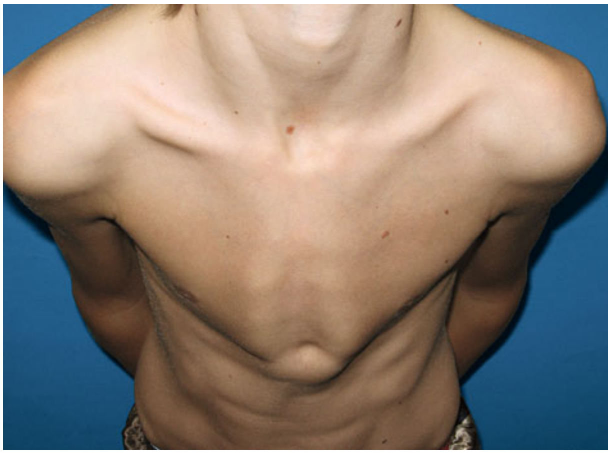

Fig. 10. 11-Year-old male with bouncing bilateral pectus carinatum ridges but only minor sternum protrusion

estimated at 1:1000 to 1:10,000. The true incidence at birth and prevalence within the population may only be derived from studies or documentations with patients that seek a physician for the purpose of information or treatment. Pectus carinatum is approximately seven times less frequent than pectus excavatum, it occurs four times more often in males than in females [9]. Also this overall ratio may be falsified by the presence of female breast volume that, in contrast to a funnel chest deformity, may conceal prominences to a major extent, so that they remain unrecognized from others than the patient, who solely may perceive the deformity by palpation.

A majority of deformities may not yet be recognized at birth and early childhood due to a prominent abdominal belly, whereas many adolescent and adult patients during and after body maturation follow coping strategies to avoid any exposure of their upper body, thus the deformity remains generally unrecognized. As the pectus carinatum deformity usually is not affected with cardiopulmonal malfunction or impairment, many patients do not complain, except those who suffer from such an aesthetic stain or by clothing unconcealable protrusion. Usually symptoms are infrequent during early childhood, thereafter in most patients remain vague and may vary from retarded growth, exertion dyspnea, or chronic dyspnea to asthmatic attacks and palpitation [6]. Such symptoms particularly occur in few patients who develop chondromanubrial prominence at early age. Many adolescent children, however with severe pectus carinatum deformities indicate to parents and physicians that they have increasingly severe shortness of breath, often with only mild exercise, reduced stamina and endurance with activity, and exercise-induced wheezing. Respiratory excursions of the thorax are considerably reduced as a result of the increase in fixed anterior-to-posterior chest diameter and increased residual lung volume, which results in tachypnea and compensatory diaphragmatic respiratory movements [5]. However, the patients usually complain more about the appearance of their chest rather than any functional difficulties [3]. The only functional problem that apparently is cumbersome and annoying with regard to the inelastic protrusion is pain while lying face down at solid bases.

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2.3.3 Pectus arcuatum

Anton H. Schwabegger

The expression of “pectus arcuatum” is not well known or used in the medical literature [2, 5] that is why the expression is rarely and unprecisely used for a variety of mixed malformations including asymmetric pectus excavatum or pectus carinatum deformities. It describes a wave-like deformity and is derived from the latin word

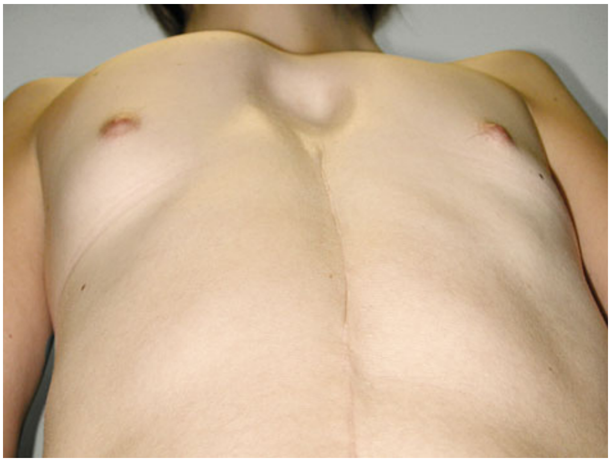

Fig. 1. Pectus arcuatum deformity containing excavatum and arcuatum features, leading to a wave-shaped curvature along the longitudinal axis

Fig. 3. Transversally orientated curvature of the anterior thoracic wall with depression at the left side and carinatum deformity at the right side

Fig. 2. Severe longitudinally orientated pectus arcuatum deformity with asymmetry (mixed deformity) in a tall adolescent with neurofibromatosis disseminata (Recklinghausen)

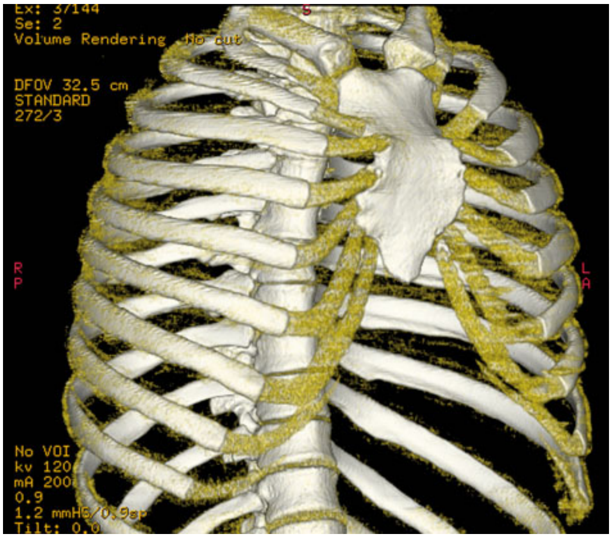

Fig. 4. Volume rendering CT scan of a pouter pigeon breast with premature ossification and significant protrusion at the angle of Louis, the manubrio–sternal transition zone

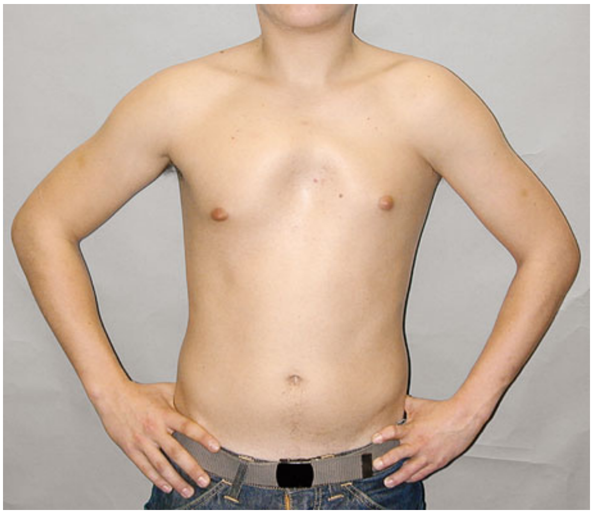

Fig. 5. Pectus arcuatum deformity in a patient with poland syndrome at the left side. Notice hypoplastic pectoralis major muscle and left arm including brachydactylia

Fig. 7. Young adult female with pouter pigeon breast and concomitant dextrocardia

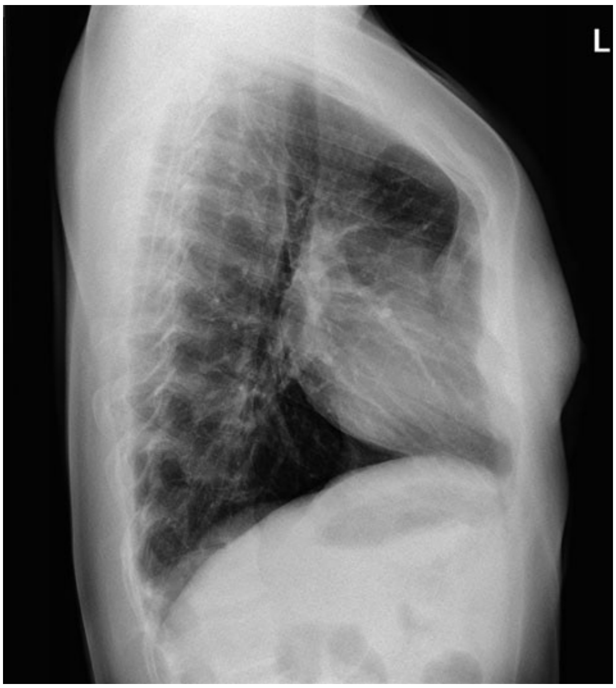

Fig. 6. Lateral radiogram depicts a prominent angle of Louis in this adolescent female with a pouter pigeon deformity

"arcus" meaning an arch or a curvature. To our understanding, the expression should only be used, if a mixed deformity contains excavatum and carinatum features, either along a longitudinal (Figs. 1 and 2) or along a transversal axis (Fig. 3), resulting in a multiplanar curvature of the sternum and adjacent ribs (Fig. 4). Such deformity as well as other malformations at the anterior thoracic wall may concomitantly also be present in a Poland syndrome (Fig. 5).

Robicsek in 1979 and Fokin in 2000 described a deformity, which was classified as pouter pigeon breast [1, 3, 6]. It contains a protrusion at the manubrio–sternal junction with premature ossification at the angle of Louis and protrusion of the second to the fifth rib cartilages (Figs. 4 and 6). The deformity in up to 30% may be associated with an excavatum deformity at the lower part of the sternum. Fokin emphasized that such a deformity with apparently absent cardiothoracic compression nevertheless may present with cardiovascular abnormalities (Fig. 7), the most common being a ventricular septum defect. All patients with a pouter pigeon deformity therefore should undergo echocardiogram to detect such intrathoracic malformations timely [1, 3].

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2.3.4 Poland syndrome

Fazel Fatah, Anton H. Schwabegger

Alfred Poland in 1840 noticed the partial absence of the left pectoralis major (PM) muscle in a cadaver during autopsy dissection and described his findings in the Guy's Hospital Reports [27]. He also noticed an ipsilateral hand deformity in the same cadaver but without making an association between the two. Clarkson in 1962, who was a plastic and hand surgeon at Guy's Hospital, noticed the association between the chest and hand deformities in some of his patients, and upon discovering Poland's earlier report, he published his findings in the same hospital journal and called the association of the two deformities Poland's syndrome [11]. This eponym has stirred some controversy over the years and indeed it is even disputed whether Poland was the first to describe the deformity in the medical literature [12, 24, 29]. However, nowadays Poland's syndrome, as an eponym, describes a wide range of possible combinations of congenital deformities of varying severity that can appear in the same patient and exceeds way beyond the combination of pectoral aplasia and dysdactylia [8].

2.3.4.1 What is involved in Poland's syndrome?

Although many types of common breast asymmetries and some chest wall deformities are attributed to Poland's syndrome, the congenital deformity itself is relatively rare and it is believed to result from a vascular event during the critical sixth week of gestation with hypoplasia of the subclavian artery causing musculoskeletal malformations [4, 7, 15, 18]. Although there are sporadic reports of familial incidence, no genetic association seems to exist [9, 14, 22, 31, 33].

Poland's syndrome is a unilateral congenital deformity that affects males and females. A ratio of 3:1 has been quoted [23], however, in clinical practice, more females seek surgical correction due to the effect and the impact the chest deformity has on the female patients. The true incidence of the deformity in the newborns is difficult to ascertain as many minor chest wall deformities at birth are not diagnosed correctly, not infrequently breast asymmetry of size is referred to as Poland's syndrome. Significantly, a very wide range of incidence has been reported in the literature, from 1 in 7,000 to 1 in 100,000 live births [9, 16, 23, 30]. In a review of 95,000 mammograms, Aznar et al. [3] looked at those patients who had breast asymmetry and recalled them for physical examination. Only six patients were found to have a combination of breast hypoplasia and partial absence of PM muscle, one of which was a male patient.

Partial absence, sternocostal head, or complete absence, both sternocostal and clavicular heads, of PM muscle and associated breast deformity is diagnostic of Poland's syndrome. The breast deformity ranges from mild asymmetry of shape (Fig. 1a–e) and size including the nipple areola complex (NAC) to severe hypoplasia that becomes more obvious as the patient reaches adulthood, or even aplasia including athelia or polythelia (Fig. 2) on the other hand. Part of the chest wall deformity is caused by a varying degree of hypoplasia of subcutaneous tissues, the superficial fascial system, and in severe cases the skin looks as though tethered to the underlying chest wall structures that can make dissection of a subcutaneous plane very difficult, particularly in the absence of the underlying ribs and intercostal muscles (Fig. 3a, b). This deficiency of subcutaneous tissues contributes to the overall asymmetry and often overlooked during the surgical correction. The oblique anterior axillary fold is missing and the skin in this area can be very tight (Fig. 1c, f) which makes the surgical recreation of the fold very difficult. In some patients there is a tight fibrous band across the axilla (Fig. 1d), crossing from the chest to the arm, which is palpable and may cause some restriction of the shoulder movement in some patients, therefore, require division. Axillary fat pad is normally underdeveloped and the hair density of the axilla is reduced (Fig. 1c). All the ipsilateral muscles with attachments to the thorax can be involved in the deformity and may affect the planning of surgical correction. The more commonly reported other affected muscles, hypoplastic or absent in some, include pectoralis minor, serratus

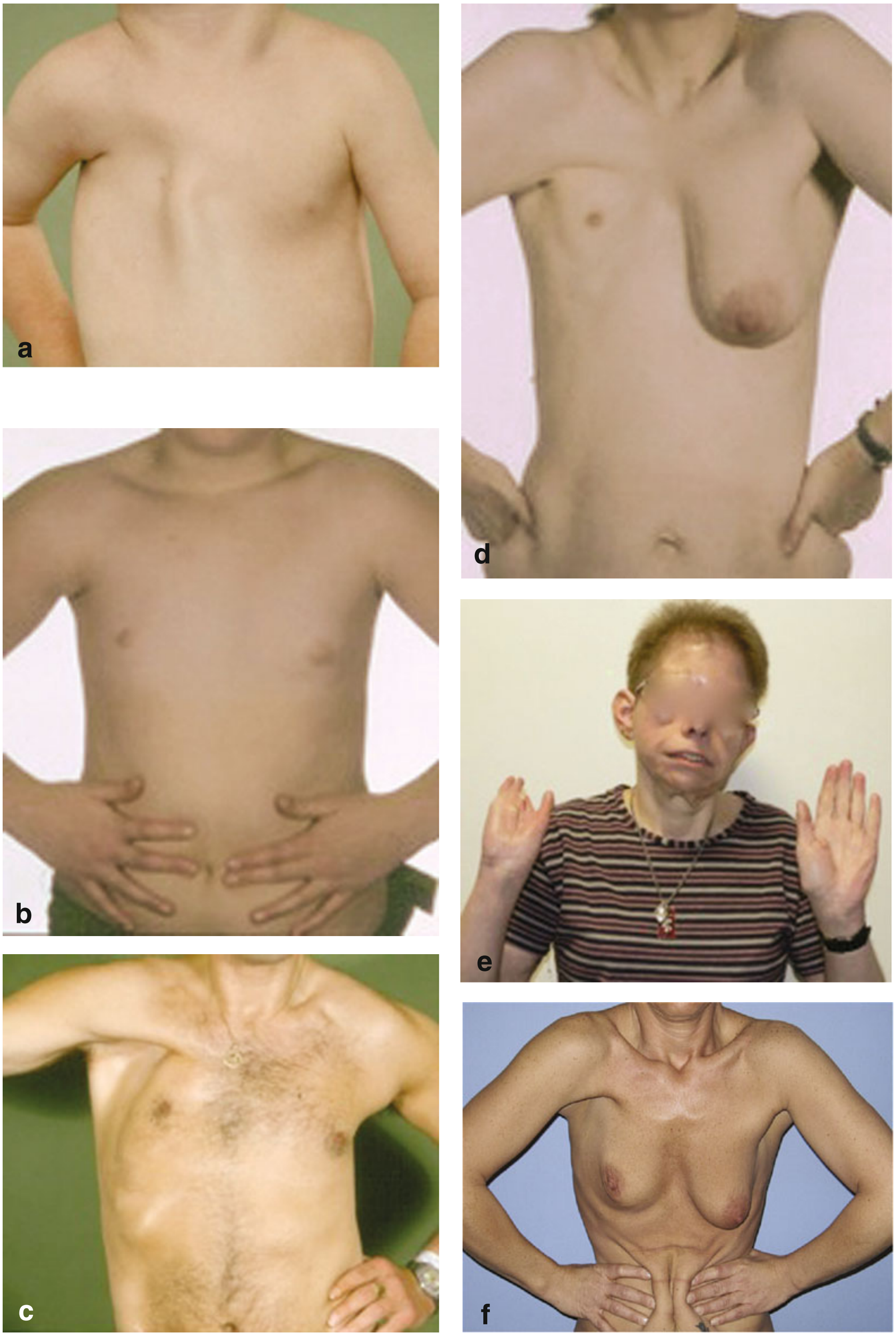
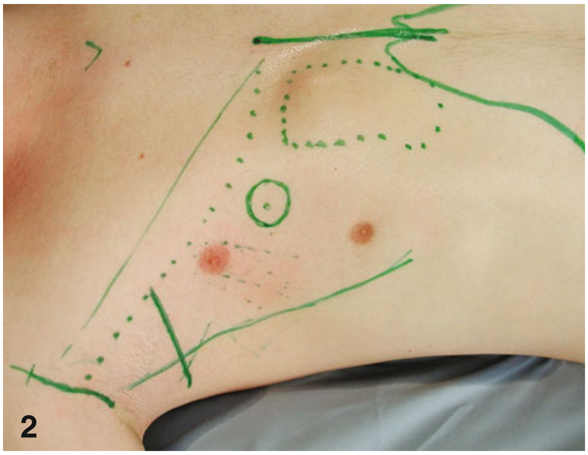
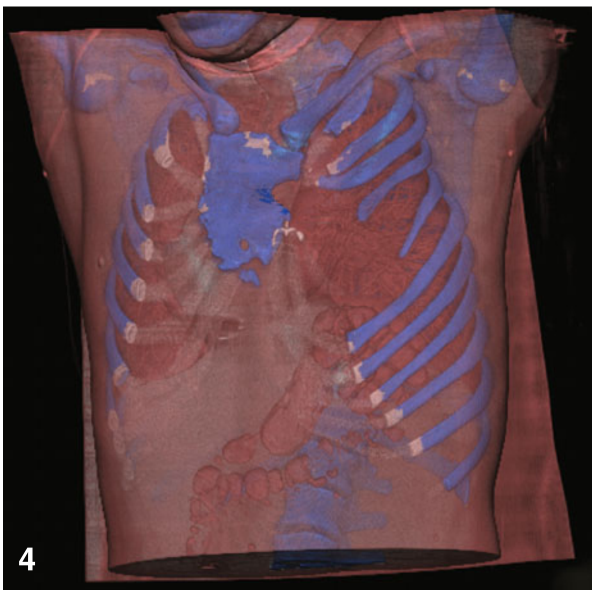


Fig. 1a–d. Different levels of chest wall deformity and asymmetry due to Poland's syndrome depending on the level of involvement of the different tissues in the deformity. **c** Also shows associated scoliosis. **e** Shows associated deformities of the ipsilateral upper limb and bilateral Moebius syndrome (patient did not wish her chest wall deformity included in the picture). **f** Hypoplastic pectoralis major muscle and breast at the right side

anterior, intercostals (normally associated with absent ribs), latissimus dorsi (LD), external oblique, rectus abdominis, supraspinatus, infraspinatus, and deltoid muscles [5, 10]. Even partial absence of the diaphragm with thoracic migration of the liver has been reported [29]. The rib cage is often involved in the deformity, ranging from minor asymmetry of shape to severe malformation including missing sections of multiple ribs and sternal deformities (Figs. 4 and 5). Ipsilateral-associated hand deformities such as symbrachydactyly (Fig. 6a, b), microdactyly (Fig. 7) and hypoplasia of the hand (Fig. 8) or of the whole limb (Fig. 9) are other recognized features of the syndrome but not always present in the same patient. Also there is no





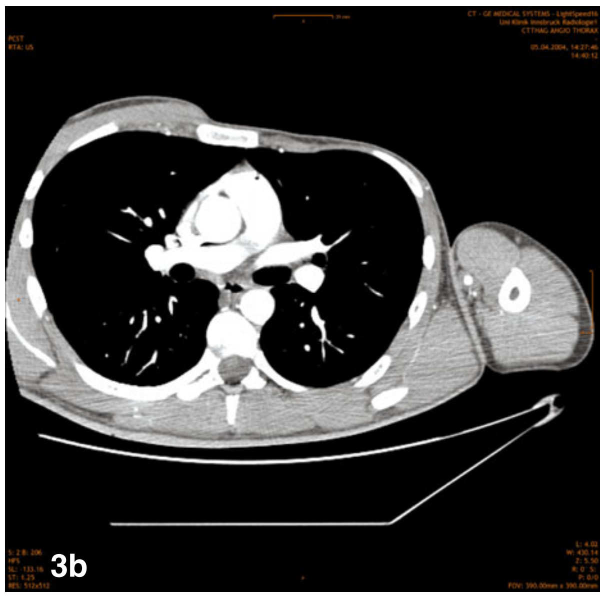


Fig. 2. A 25-year-old male with absent sternal and costal portion of the pectoralis major muscle, rib hump deformity (dotted rectangular line), minor malrotation of the sternum and polythelia. The natural position of the NAC is marked with a circle

Fig. 3a. MRI scan showing absent pectoralis major muscle beneath a slightly hypoplastic breast at the right side. **b** MRI scan in a male with absent pectoralis major muscle and underdevelopment of ribs. Notice also under-developed subcutaneous tissue with only a thin tissue layer covering the pleura

Fig. 4. Volume rendering three-dimensional CT scan of a 24 years old male with hypoplastic ribs at the left side, partially missing rib cartilages and misfigured sternum (Courtesy of Dr. J. Jeschke, Hospital of Mercy Sisters, Linz, Austria)

Fig. 5. Pectus arcuatum deformity in a 16 years old male. Absent pectoralis major muscle and malposition of the NAC, slightly hypoplastic upper arm at the left side

Fig. 6a. A 6 months old female with symbrachdaktyly with Poland syndrome. **b** Same patient 17 years later after several hand surgery interventions for separation of the digits to improve function

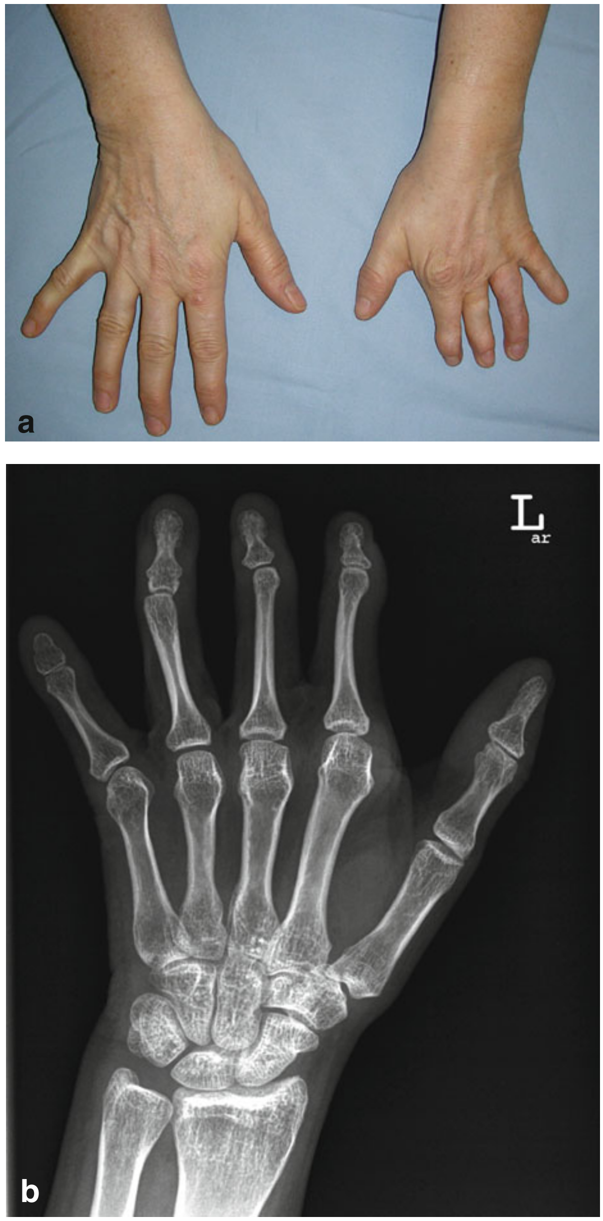


Fig. 7a. A 46-year-old female with microdaktyly at her left hand and Poland syndrome. **b** X-ray of the same patient

direct relation between the severity of hand deformity and that of the chest [17]. Different classifications for associated limb and hand deformities in Poland's syndrome have been suggested, depending on the severity of the condition [2, 19]. Scoliosis is often noticed in Poland's syndrome, which may further complicate the overall appearance and the treatment of the chest wall deformity. Other recognized associated congenital abnormalities include Moebius syndrome [1, 22, 28, 32],

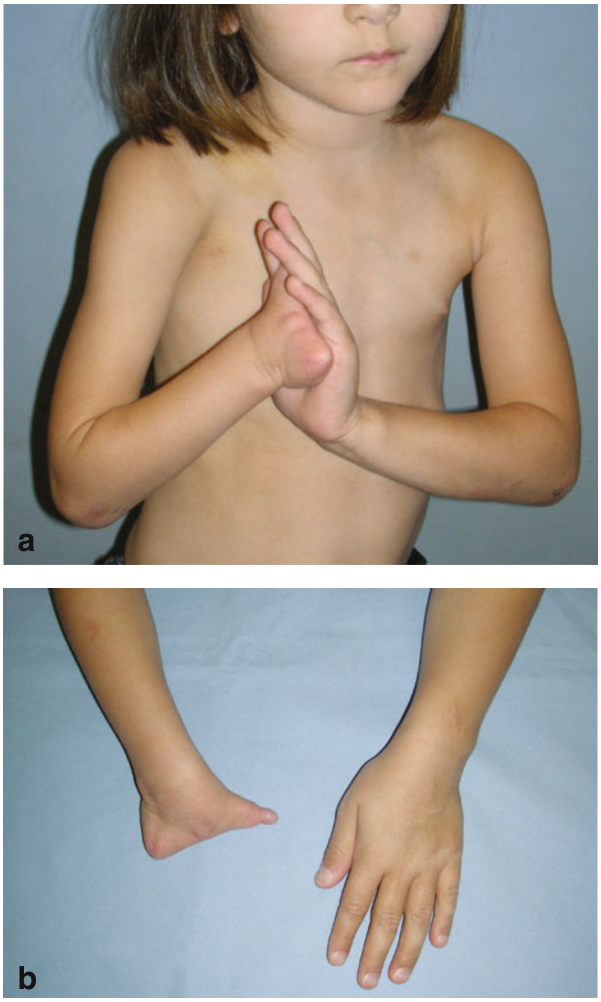


Fig. 8a, b. Severe hypoplastic left hand in a 6 years old female with Poland syndrome

Fig. 9. A 13-year-old male with Poland syndrome and hypoplastic right arm

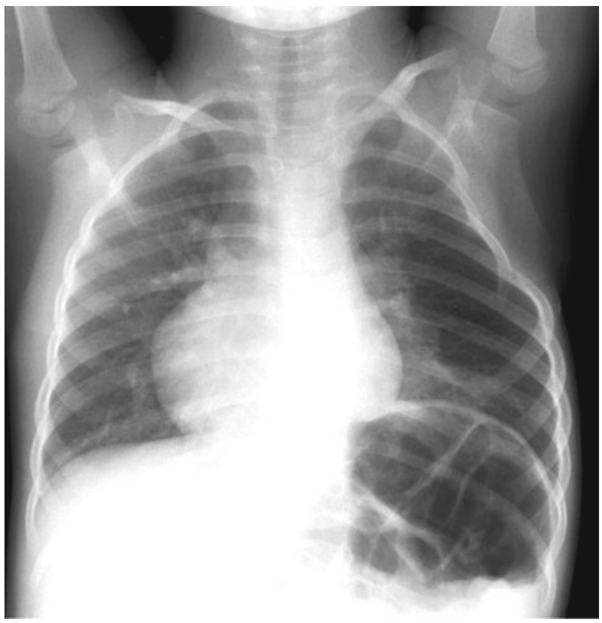


Fig. 10. A 10-year-old female with dextrocardia and hypoplastic thoracic cage at the left side, partially absent anterior parts of the third and fourth rib

contra-lateral peripheral facial palsy [34], growth hormone deficiency [21], megacalycosis [9], dextrocardia (Fig. 10) [6], optic nerve abnormalities [26] and facio-auriculo-vertebral dysplasia [13].

The entity of Poland's syndrome with absent female breast was named "Amazone" syndrome by Mühlbauer in 1977 [25], but represents a selected description of a segment out of the whole complexity of variable expressions of Poland's syndrome. Mühlbauer stated that simultaneous damage to the extremity bud and the close-by lacteal mound during embryonic development might result in a concomitant breast aplasia (Fig. 1d). However, a majority of all of the female patients with Poland's syndrome shows variable expressions of breast hypoplasia down to aplasia at the site of muscular and skeletal deformities, while the minor forms often are misdiagnosed as simple breast asymmetry. On the other hand one might argue that all kinds of unilateral breast hypoplasia represent minor variants of Poland's syndrome in a mitigated form, which could not be confirmed in a radiological and ensuing clinical study [3]. However, the complexity and variability of expression of these deformities deserve further attention and accurate classification with regard to diagnosis and ensuing therapeutic options.

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2.3.5 Cleft sternum and other anomalies

Barbara Del Frari

Sternal cleft, described as early as 1772 by Sandifort, 1798 by Wilson, 1818 by Weese, is a rare congenital anomaly [1, 13, 15]. It results from failure of fusion of the two lateral mesodermal sternal bars by the eighth week of gestation. The development of the rest of the sternum and its association with the ribs laterally usually remains normal.

Depending on the degree of separation, there exist complete and incomplete forms. Both the upper and the lower parts of the sternum may be involved, or there may be as the most rare expression, a complete sternal non-union. Superior clefts are more frequent than inferior ones, but isolated central clefts are extremely rare.

Clinically, three groups of expression of sternal clefts can be identified:

1. *Cleft sternum without associated anomalies*. Because the fusion during embryonal development starts from above, the defects reach either through the entire sternum or only the lower part is affected. On the basis of the arrangement of the sternal defect, bifid sternums can be either V or U shaped [7].
2. *True ectopia cordis*. The heart is elevated anterior to the chest wall along with some degree of sternal cleft.
3. *Cantrell's pentalogy*. The anomalous association of defects of the midline, at the supraumbilical abdominal wall, the lower sternum, the anterior diaphragm, the pericardium, with congenital intracardiac defects since 1958 has come to be called after Cantrell [4].

True ectopia cordis and Cantrell's pentalogy are also summarized as disorders of ventral closure [17].

2.3.5.1 Isolated cleft sternum

This is a very rare congenital deformity, with only about 100 instances being reported in the literature [7]. In one series published, isolated cleft sternum occurred in 0.15% of 5,182 patients referred for management of chest malformations [1]. A female predominance has been reported but familial expression seems to be nil. The sternal cleft either present at the superior or inferior part (Figs. 1 and 2) of the sternum frequently is associated with supraumbilical raphe and disturbances of abdominal wall midline fusion [10, 11].

Although prenatal diagnosis of a sternal cleft can be made by ultrasonography, isolated sternal clefts generally first present at delivery [16]. There is always normal skin and soft tissues covering the bony defect through which the pulsation of the heart and respiratory movements of the lung are visible. These children are generally asymptomatic, but dyspnea and recurrent pulmonary infections can occur. Isolated sternal clefts are rarely associated with cardiac defects and are often associated with vascular mal-

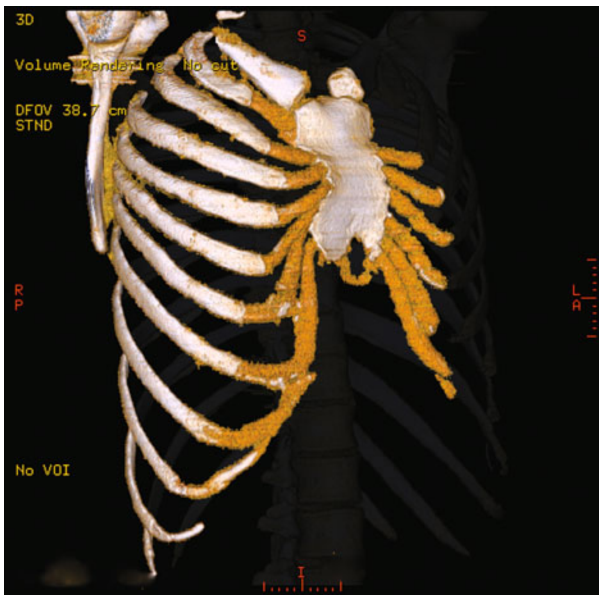

Fig. 1. Sternal foramen at the lower end of the sternal corpus in a 16-year-old female. The potential presence of such or a similar foramen might cause troubles during diagnostic sternal puncture with the risk of immediate perforation into intrathoracal structures, but apart from that remains without symptoms

Fig. 2. A 14-year-old male apparently with incomplete and narrow sternum cleft at the lower portion of the sternum corpus, associated with pectus carinatum deformity. Apart from an esthetic issue without any somatic complaints

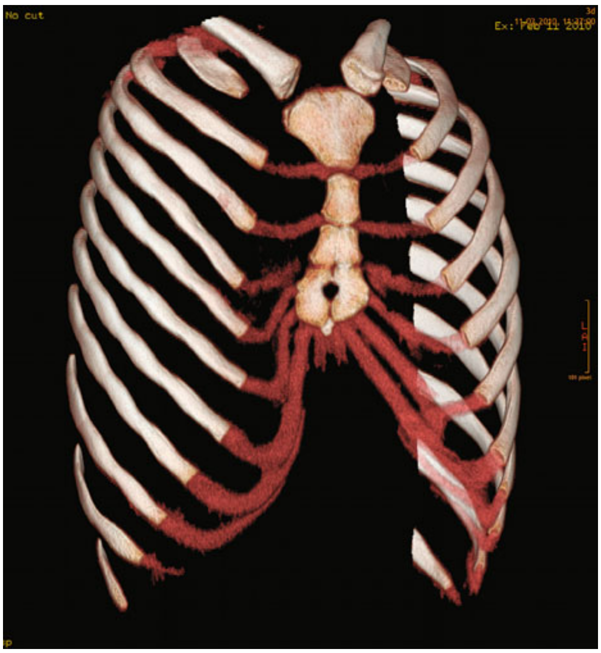

Fig. 3. Incomplete cleft sternum associated with a pectus arcuatum deformity, dextrocardia and massive vertebral scoliosis in a 20-year-old female

formations (e.g., craniofacial hemangiomas). Sternal foramen (Fig. 3) usually causes no complaints for the patient, but awareness about its potential presence is important in the cases of punctures indicated by medical reasons. Because of the absence of particular functional disorders these patients have long-term survival rates like the healthy normal population.

2.3.5.2 Thoracic (true) ectopia cordis

Ectopia cordis is a congenital anomaly defined as a congenital malposition of the heart, partially or completely (Fig. 4) outside and anterior to the thoracic cage [6]. This condition is associated with a high mortality rate, largely because of the associated intracardiac defects [10].
The diagnosis is made either prenatally by ultrasound examination or immediately after birth with the observation of the heart being at least partly out of the chest [13, 20].
Ectopia cordis is sub-classified into five types based on the location of the heart: cervical, cervio-thoracic, thoracic and more commonly thoraco-abdominal, furthermore abdominal ectopia [13]. Cervical and cervicothoracic ectopia cordis are uniformly lethal. The thoraco-abdominal ectopia cordis is more amenable to surgery and is often associated with the pentalogy of Cantrell [4]. The thoracic type also carries a high mortality risk. Abdominal ectopia cordis with displacement of the heart into the abdomen often needs no specific intervention for survival [12, 13]. Thoracic (true) ectopia cordis with displacement across the chest wall occurs in about 38% of all these patients [14].
Thoracic ectopia cordis necessarily in association with sternal cleft or absence of sternum [12] is extremely rare with a reported incidence of 5.5 per 8 million live births [10]. In partial thoracic ectopia cordis the heart can often be seen pulsating through the skin. In complete thoracic ectopia cordis the

Fig. 4. Newborn with a wide cleft sternum and true ectopia cordis (Picture with courtesy of Prof. J. Hager MD, Department of Pediatric Surgery, Medical University Innsbruck)

naked heart is displaced outside the thoracic cavity without pericardial coverage [2]. These patients have a poorer survival [18] also because cardiac defects are present in 80% of these patients. The most common defects are ventricular septal defects, atrial septal defects (53%), tetralogy of Fallot (20%), and hypoplastic lungs [2].

2.3.5.3 Pentalogy of Cantrell

Cantrell in 1958 described a pentalogy characterized by a midline supraumbilical abdominal wall defect (omphalocele), a defect of the lower part of the sternum, a deficiency of the anterior diaphragm, a defect of the diaphragmatic pericardium, and a congenital heart malformation [4, 13].
The estimated incidence of this malformation is about 1 per 65,000 live births with a slight male predominance [8]. The syndrome can be diagnosed by ultrasound examination in utero already but has an extremely poor prognosis. Associated malformations might include midline craniofacial malformations like cleft lip and encephalocele [19]. Abdominal wall defects occur in 74.5% of these patients, sternal defects in 59.4%, and diaphragmatic defects in 56.8% [8].

2.3.5.4 Other disorders of the rib cage

The asphyxiating thoracic dystrophy (JATD), also known as Jeune syndrome is a rare congenital disorder with a prevalence in one study of 2.6 per 100,000 [3]. It is defined as "a multi-system autosomal recessive disorder associated with characteristic skeletal dysplasia and variable renal, hepatic, pancreatic, and retinal abnormalities" [9]. Additional bone anomalies include irregular curvature of the clavicles, shortening of ulna, radius, tibia, fibula, and a small irregular pelvis [13, 21]. Also polydactyly of hands and feet is often present. Usually the main issue is respiratory failure secondary to a dysplastic rib cage. Short horizontal ribs limit lateral expansion of the small chest cavity thus ventilation is almost exclusively diaphragmatic. The condition of the underlying lungs can range from normal to severely hypoplastic, with associated pulmonary hypertension.
Ultrasound has been used to diagnose JATD in the prenatal period as early as 14 weeks gestation by measurement of limb length or disparity of fetal thoracic and abdominal circumferences [5, 13].

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2.3.6 Syndromal, mixed, and other deformities

Barbara Del Frari, Anton H. Schwabegger

A congenital chest wall deformity can be an isolated malformation or dysmorphic feature or only one symptom of a genetic syndrome [2]. So far there are only rare syndromal deformities of the anterior chest wall known, exemplarily the Poland syndrome (Chapter 2.3.4) as an own entity, with

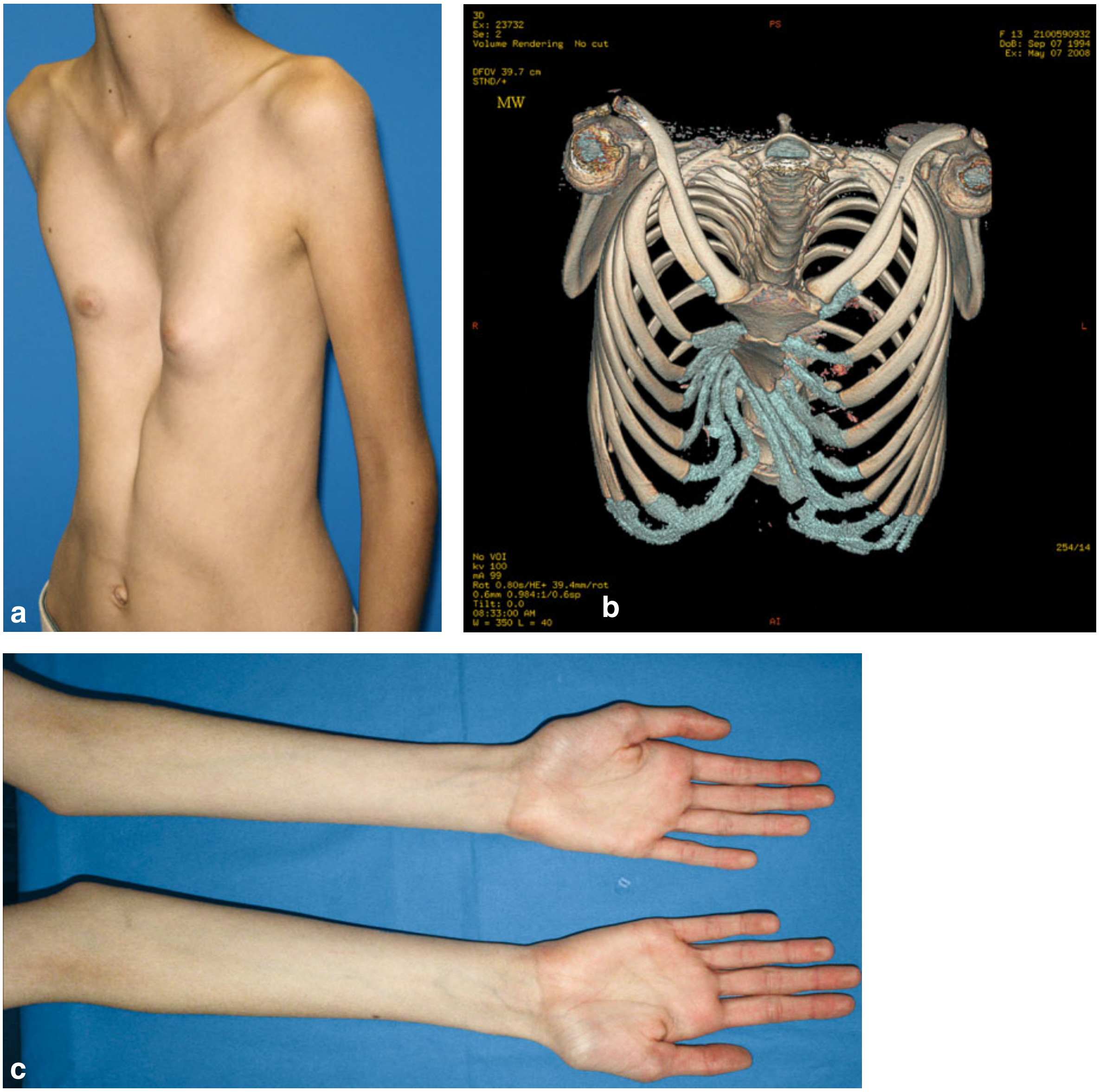

Fig. 1. Extensive pectus excavatum deformity (**a**) in a girl with Marfan syndrome, apparent sternum malrotation (**b**), and arachnodactylia (**c**)

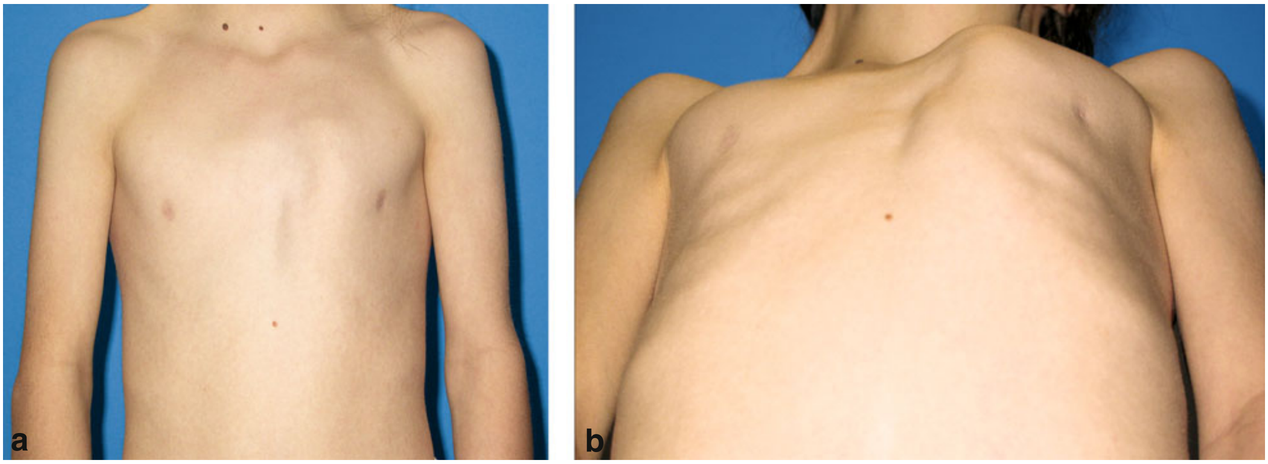

Fig. 2. Nine-year-old female with Ulrich-Turner syndrome, visible with pterygium colli (**a**) and keel chest deformity (**b**)

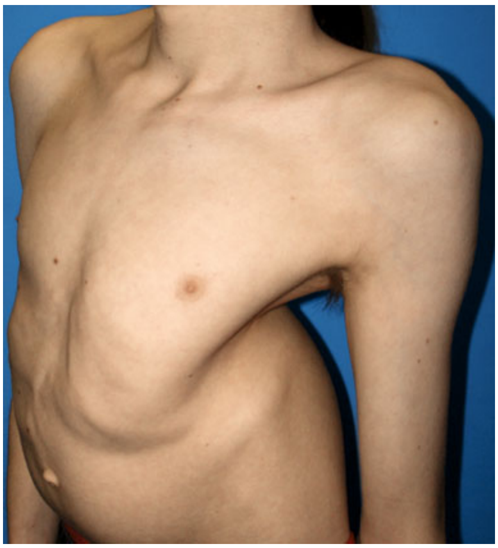

Fig. 3. 19-Year-old male with severe congenital scoliosis, resulting in rib humps and multiple distorted ribs

different extents of phenotypic expressions or syndromal cleft sternum as Cantrell's pentalogy (Chapter 2.3.6). Nevertheless several syndromes show coincidence with chest wall deformities. First of all the Marfan syndrome (Fig. 1a–c) and the Noonan syndrome (Chapter 2.2) may be associated with thoracal anomalies, as well as Ulrich-Turner syndrome (Fig. 2a and b). However, chest wall anom-

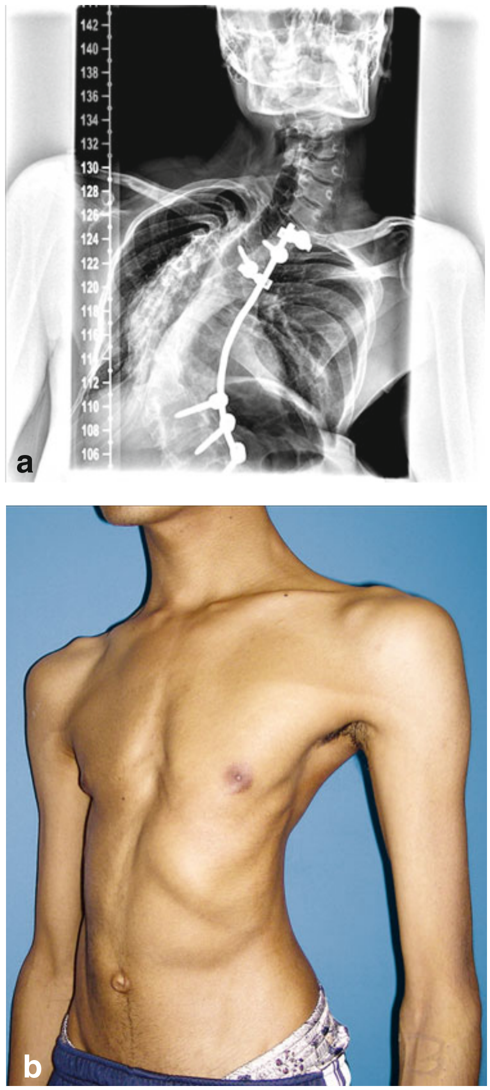

Fig. 4. 20-Year-old male with severe congenital scoliosis already surgically corrected (**a**), without the need for surgery at the deformed ribs (**b**)

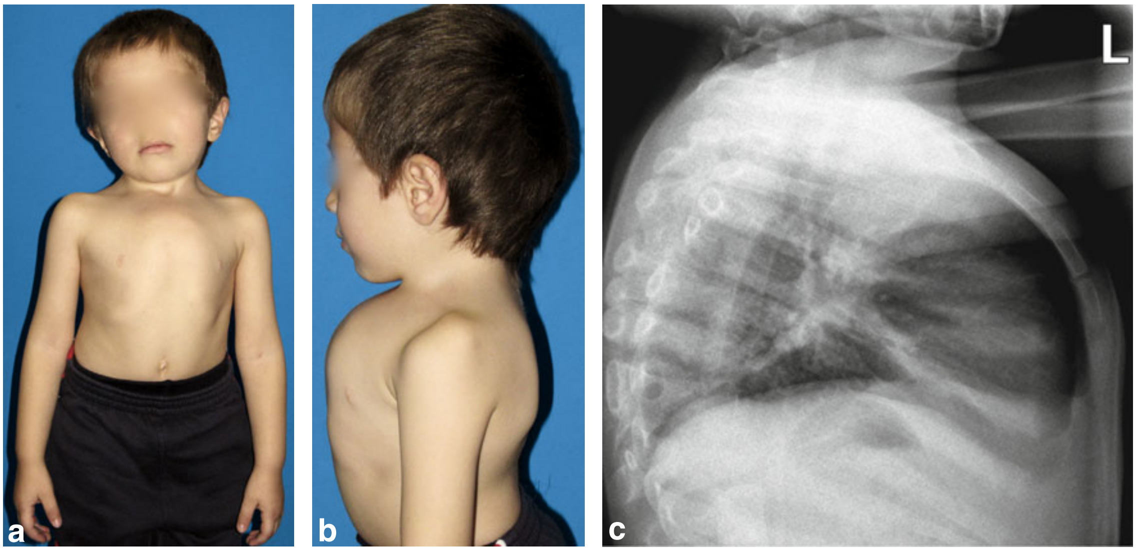

Fig. 5. Four-year-old boy with spondylocostal dysostosis type I, scoliosis and nanosomia (**a**, **b**) with resulting barrel chest deformity (**c**)

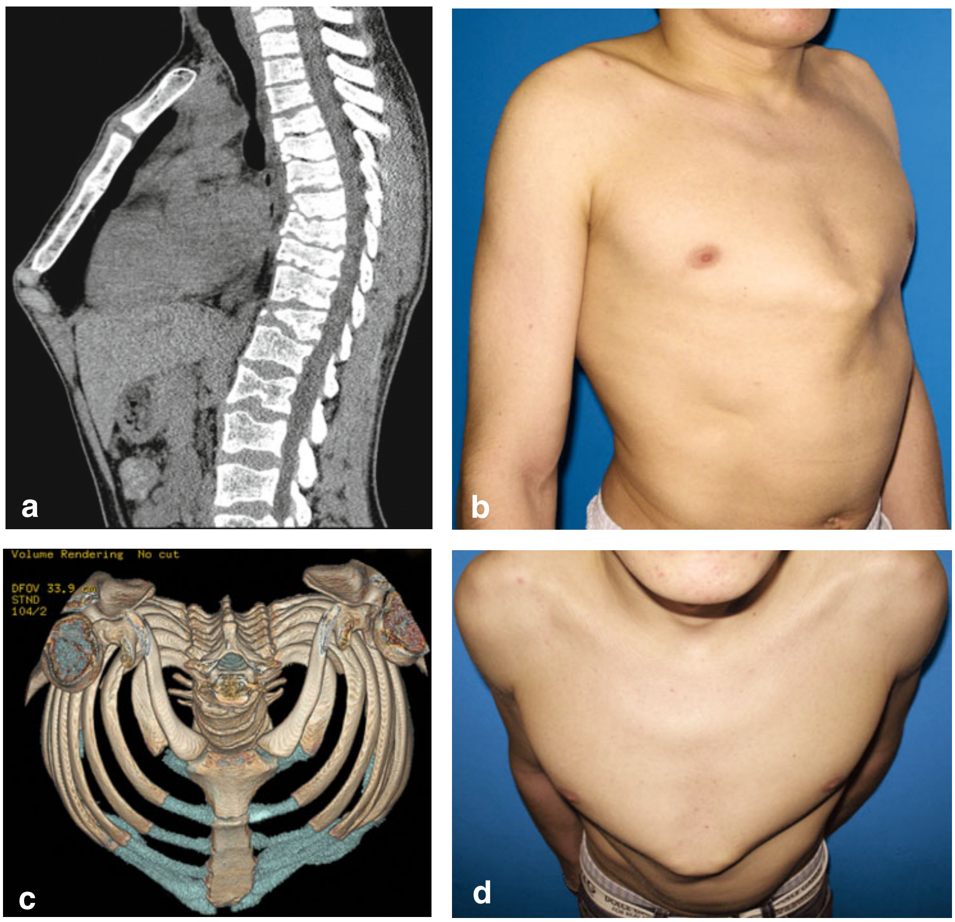

Fig. 6. Median scan (**a**) of keel chest deformity in a male with multiple epiphyseal dysplasia, notice the multiple dysplastic vertebral bodies which result in nanosomia and extensive keel chest deformity (**b**, **c**, **d**)

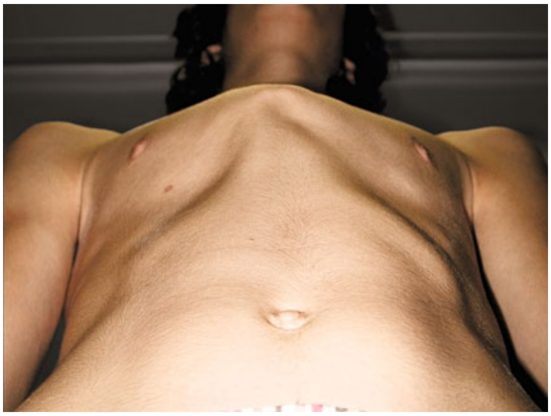

Fig. 7. Mixed deformity with leading feature of keel chest but bilateral rib depressions at the infrapectoral regions

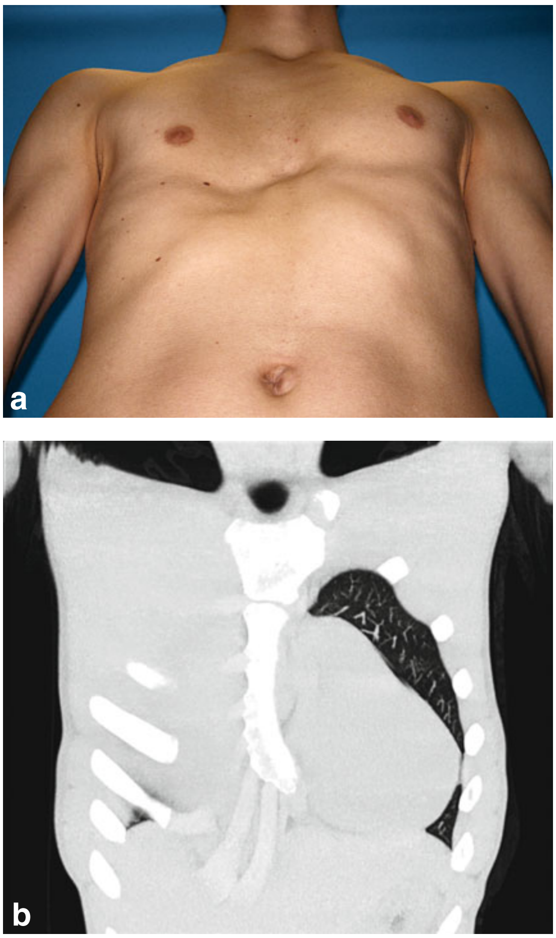

Fig. 8. Deformity with leading feature of flat asymmetric pectus excavatum and sternum malrotation (**a**). Only after radiographic imaging that depicts an additional curvature within the sternum in a frontal plane (**b**), the deformity may be classified as mixed or complex

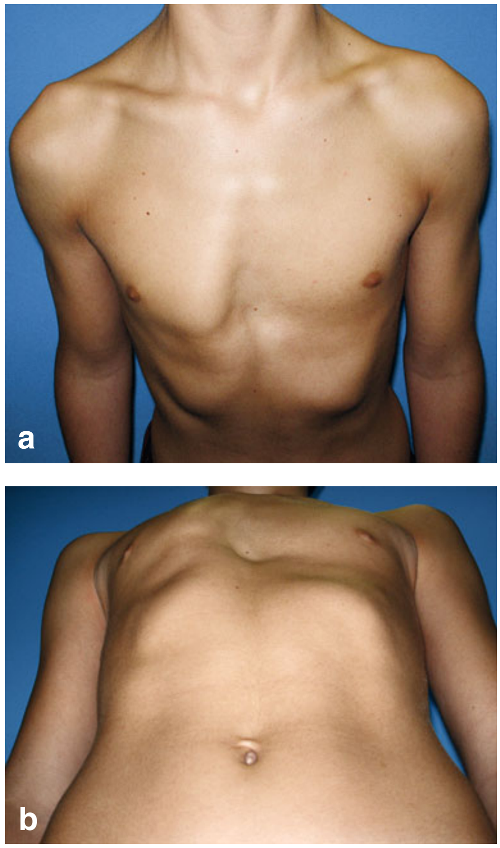

Fig. 9a and b. Mixed deformity with leading feature of asymmetry but equivalently disturbing expression of keel chest at the right parasternal side and funnel chest at the central sternal area

alies are not pathognomonic for a syndrome and in most cases they clinically represent a single entity of deformity. Nevertheless when first treating a patient having a chest wall anomaly, further examination is mandatory to exclude potential coincidences with syndromal diseases. Often patient and parents are not aware of eventually present syndromes but focus their principal attention on the apparent chest wall anomaly.

Commonly on the other hand severe congenital scoliosis of the vertebral column is associated with chest wall anomalies. There is however still lack of knowledge about the pathogenesis of thoracic anomalies going along with scoliosis, whether they are expression of a secondary reactive deformation based on compelled biomechanical forces due to heavily distorted and multiaxially shortened thorax (Figs. 3 and 4a, b) or if they are genetically contingent and accompanied with the scoliosis deformity itself (Figs. 5a–c and 6a–d).

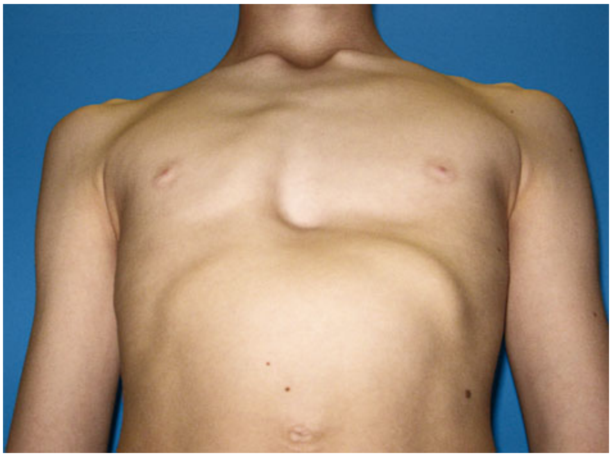

Fig. 10. Mixed deformity with leading feature of pectus excavatum at the left side and central sternal area, while only minor expression of a keel deformity is present at the right parasternal side

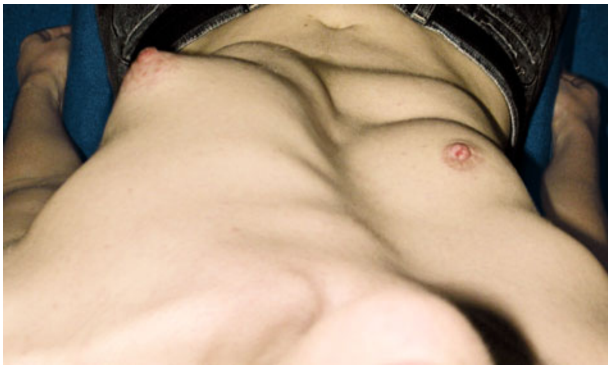

Fig. 11. Severe mixed deformity with hemithoracal hypoplasia, major twisting of sternum and breast volume asymmetry

Fig. 12. Mixed deformity with leading feature of symmetric platythorax pectus excavatum and major protrusion of the lower rib arches

Fig. 13. Assumed acquired keel chest deformity, based on Marfan syndrome and aortic surgery 2 years before the photo was taken. The transsternal access was accused for the development of the protruding deformity but cannot be verified due to unknown implications from abnormal epiphyseal growth going along with the Marfan syndrome

Among the congenital anomalies of the anterior chest wall, the complex or mixed deformities represent such cases that cannot be categorized into the classification of Willital [4] or other authors. Their basic description should contain the leading deformity with a circumscription of the additionally present anomalies accompanying it (Figs. 7–12). This is of importance as such indeed not very rare complex cases require a distinct strategic surgical access to enable a successful outcome (Chapter 8).

Acquired deformities of the anterior chest wall must be separated from the congenital forms, as they usually represent a late sequel of former surgery, whether in childhood for cardial or aortic repair (Fig. 13) or later during growth periods of the skeleton. Likewise with the scoliosis cases, the causality of deformity may not be definitely attributed to the surgical intervention at the thorax solely but rather to the coincidence of congenital cardial heart defect with congenital chest wall deformity which may be present. As such, the

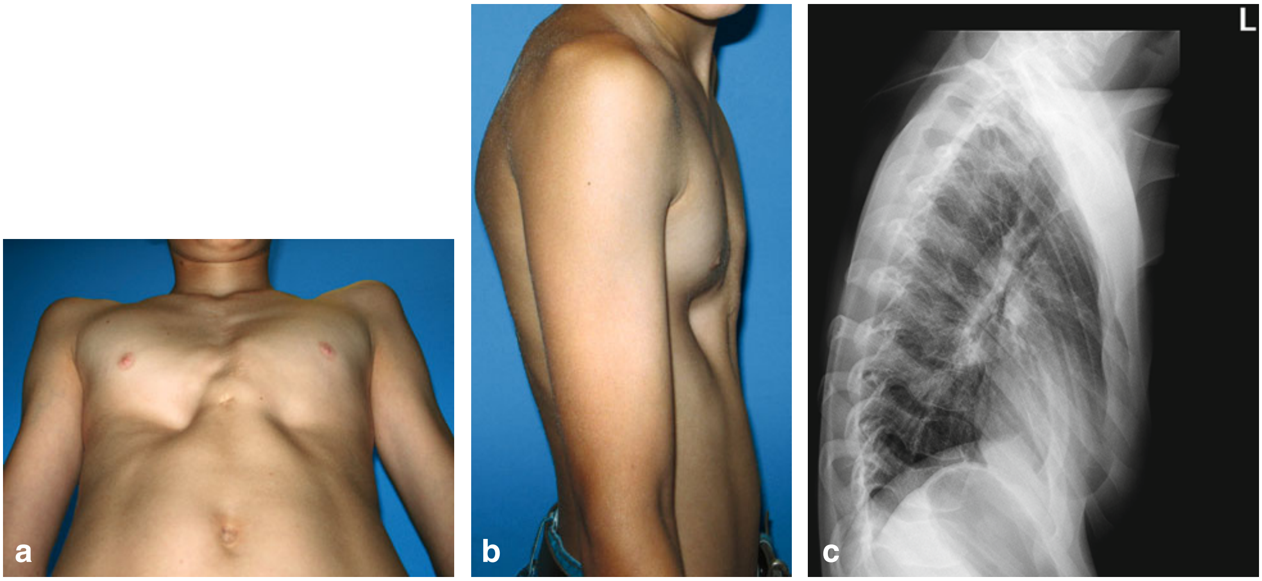

Fig. 14a–c. 14-Year-old male who underwent cardiac surgery early in the postnatal period. The deformity aggravated since then and might be based on a funnel chest deformity, its "normal" development was altered by the early surgical access. Instead of leading into a funnel deformity, it results in a presumably acquired mixed deformity with a median keel chest but still remaining extensive bilateral rib depressions

Fig. 15. Acquired deformity also described as reactive keel chest deformity after funnel chest repair with MIRPE, presumably as an effect of folding up of a steep funnel deformity by the pectus bar

sudden appearence after heart surgery will misconstrue the occurring chest wall deformity as an iatrogenic effect, but presumably is rather based on a genetic disorder. As pointed out by Haje et al. [1], growth plates at the sternum and genetically based disturbances there during growth spurts may result in sternum distortion. Likewise surgical splitting of the sternum, herein transection of epiphyseal growth plates within the sternal bones may presumably also trigger false development and reactive distortion of the sternum (Figs. 13 and 14).

A definite iatrogenic deformity may result from a pectus excavatum repair utilizing MIRPE, while reactive pectus carinatum (Fig. 15) results from further cartilage growth when a pectus bar is implanted and prohibits further elongation versus the originally present funnel deformity. Instead of that the growing cartilages push the supported sternum into a carinatum deformity [3].

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Diagnostics

3.1 Photography

Anton H. Schwabegger

"A picture is worth a thousand words."

Outcome documentation and course observation of plastic surgical interventions are described often circumstantially; however, in addition to the backup of photos and in functional reconstructions with movies, these interventions can be documented comprehensively. For decades already reconstructive surgeons rely on photography as a means for documentation of patients, assessment of results, teaching, and publications. Nowadays, with the evolution of digital depiction many tasks in storage, post-processing, and visualization are made much easier, thus standardization is a requisite for academic research [1] and to obtain informed consent from patients. Usually patients show preference to the use of non-identifiable photographs, as long as they serve for medical documentation, teaching or presentations on meetings, and congresses, whereas they do less accept distribution of pictures on the Internet [2, 3]. Therefore, also a written consent, containing the detailed purposes of taking photographs should be obtained from all patients before any photography.

A documentation of the initial situation in reconstructive or remodeling surgery serves both the planning of, at times, complex intervention and also for the preoperative information for the patients. The documentation of thoracic wall deformities by means of photography succeeds very well if certain criteria of standardized photo techniques are adhered. This standardization is an absolute requirement to enable documentation of the course of the variations through the intervention traceable and to compare the preoperative situs with the postoperative result understandably [4, 5]. Photographs such as these can be stored digitalized in a computer and can be retrieved via network in any examination room on demand during consultation or even teaching. Because in such corrective interventions at body contour often only an amelioration of the extent of the deformity is attainable and in spite of substantially best information communicated to the patient, his expectation of a perfect shape cannot be achieved. In these cases, an objective documentation by means of photography may be very helpful in the argumentation of differences of opinion or disproportionate expectations (Chapter 3.6). Herewith and during the first conversation, vivid information at hand of photographs of operated cases can already help the patient to estimate the feasibility of corrections realistically and may limit his expectations. Also the photographic presentation at hand of the results of treatment alternatives or other surgical methods can help the patient to come to a well-tolerated decision. By reason of published reports in glossy prints or semi-medical magazines many patients already reject the use of any silicone implant beforehand. Such misinformed and irritated patients then can become convinced, however at hand of already documented results of alternative interventions, that minor invasive surgery using, e.g., alloplastic material or autologous tissue may yield sometimes better results than more invasive modern interventions. Through photo documentation, the aspect of scars to be expected can be better imagined and explained.

Furthermore, an exact photographic documentation also serves for quality improvement and represents a valuable and reliable tool for quality control. Photography substantially also serves for teaching the treating nurses and students. It is a helpful instrument in training and continued education within departments as well as in interdisciplinary advanced training and on congresses. During demonstration and teaching therewith no patient with such a deformity, usually embarrassed and plagued by shame, needs to undress and expose face-to-face with other medical personnel or students. On the other hand, the presentation of photo documentation, even if comprehensively complete, cannot replace the personal conversation with the patient, its exact case history inquiry and psychological examination, but rather will supplement such a consultation ideally.

In the question and the setting of indication to any surgical intervention as a curative treatment with the costs covered by the health insurance companies, photography often also serves as a supplementary decision criterion. Finally a chronologic documentation using photography is part of the medical record and in cases of medico-legal issues with conflicts between patient and physician may serve as an objective evidence of course and results.

Standardized constants in photography of the thoracic wall deformities are:

- distance from camera to patient;
- angle of camera to patient; and
- removal of clothing, underwear, and noticeable jewellery from the torso.

Photos are made preoperatively and postoperatively up to 1 year of follow-up:

- 0° exact anteriorly (Fig. 1),
- 90° angled laterally from the right side (Fig. 2),
- 90° angled laterally from the left side (Fig. 3),
- 45° angled laterally from the right side (Fig. 4),
- 45 angled laterally from the left side (Fig. 5),
- from above (Fig. 6), and
- from beneath (Fig. 7).

In addition, specially angled photos are made in particular deformities or, e.g., asymmetry of the female breast:

- from above and beneath (Fig. 8–10) and
- torso bent over to depict the difference of volume or distortion of the breasts (Fig. 11).

In addition, specially angled photos are made in particular deformities like Poland's-syndrome to document the extent of deformity and to depict functional deficits during muscular contraction based on hypoplastic skeletal structures.

- multiple functional photos taken without and with muscular contraction, individually dependent on the extent and type of deformity (Fig. 12 and Fig. 13a–k).

Photos of the initial preoperative situation are compared with the immediate postsurgical appearance to document the immediate morphologic and aesthetic success of an intervention exactly. In the further follow-up examinations, photos are taken after suture removal, furthermore after 3, 6 and 12 months in order to observe the course of the result objectively and to recognize any possible morphologic variations as a side-effect for example due to physical growth. These photos then serve, together with chest X-rays and the measurements taken with the Thorax-caliper (Chapter 3.2) as multimodal documentation and serve to fulfil the increasing demand for quality control and quality security over a longer term. In consequence, such long-termed follow-up documentation serves for the scientific editing, with reliable and valuable verification of results and the option to facilitate peer-reviewed studies [1] and publication of meaningful and permanently successful or also less successful interventions. Particularly the depiction and description of undesirable results or long-term complications serve the progress and the selection of meaningful measures out of a multitude of most varied therapeutic measures or therapy attempts.

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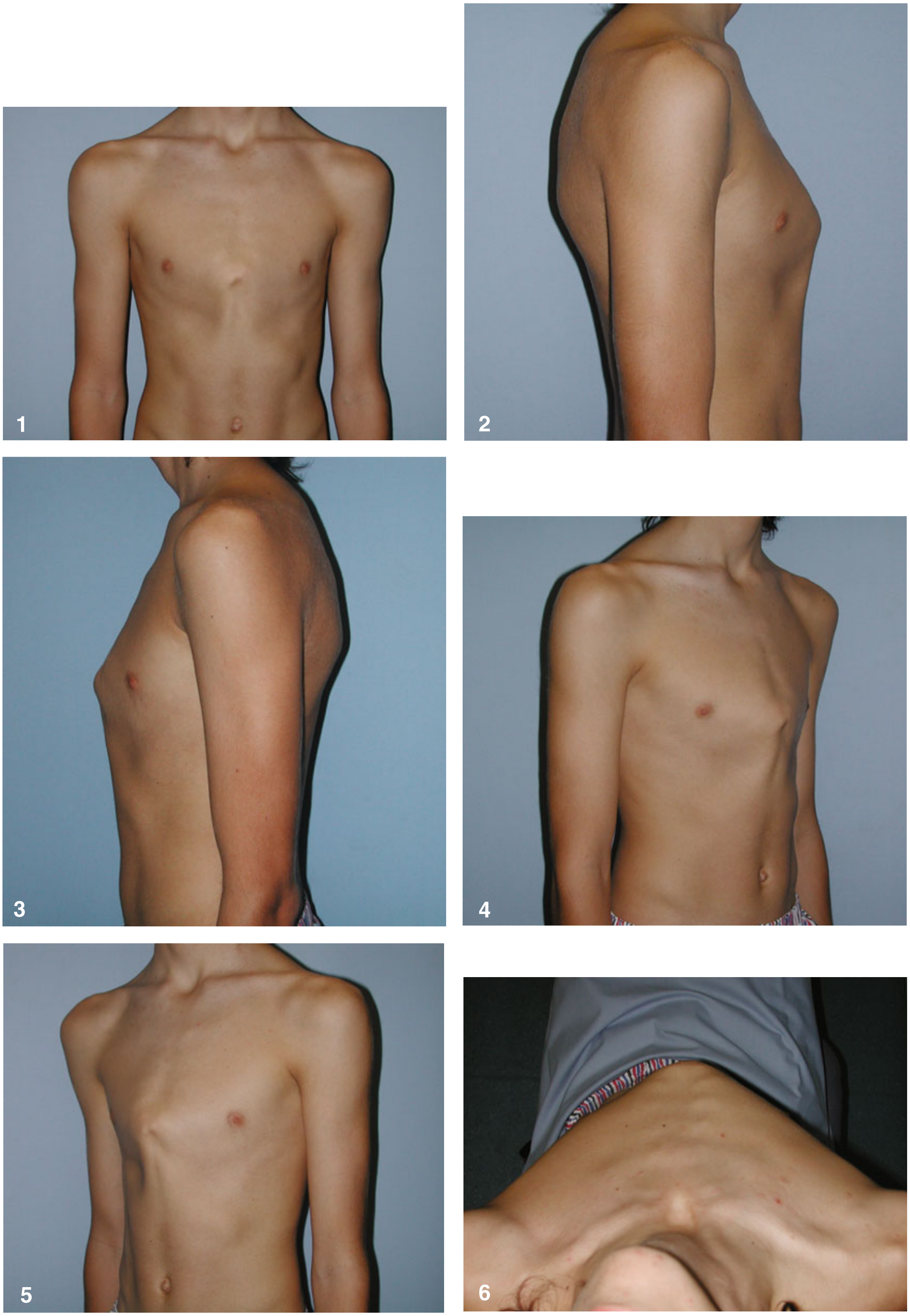


Figs. 1–6

Figs. 7–12

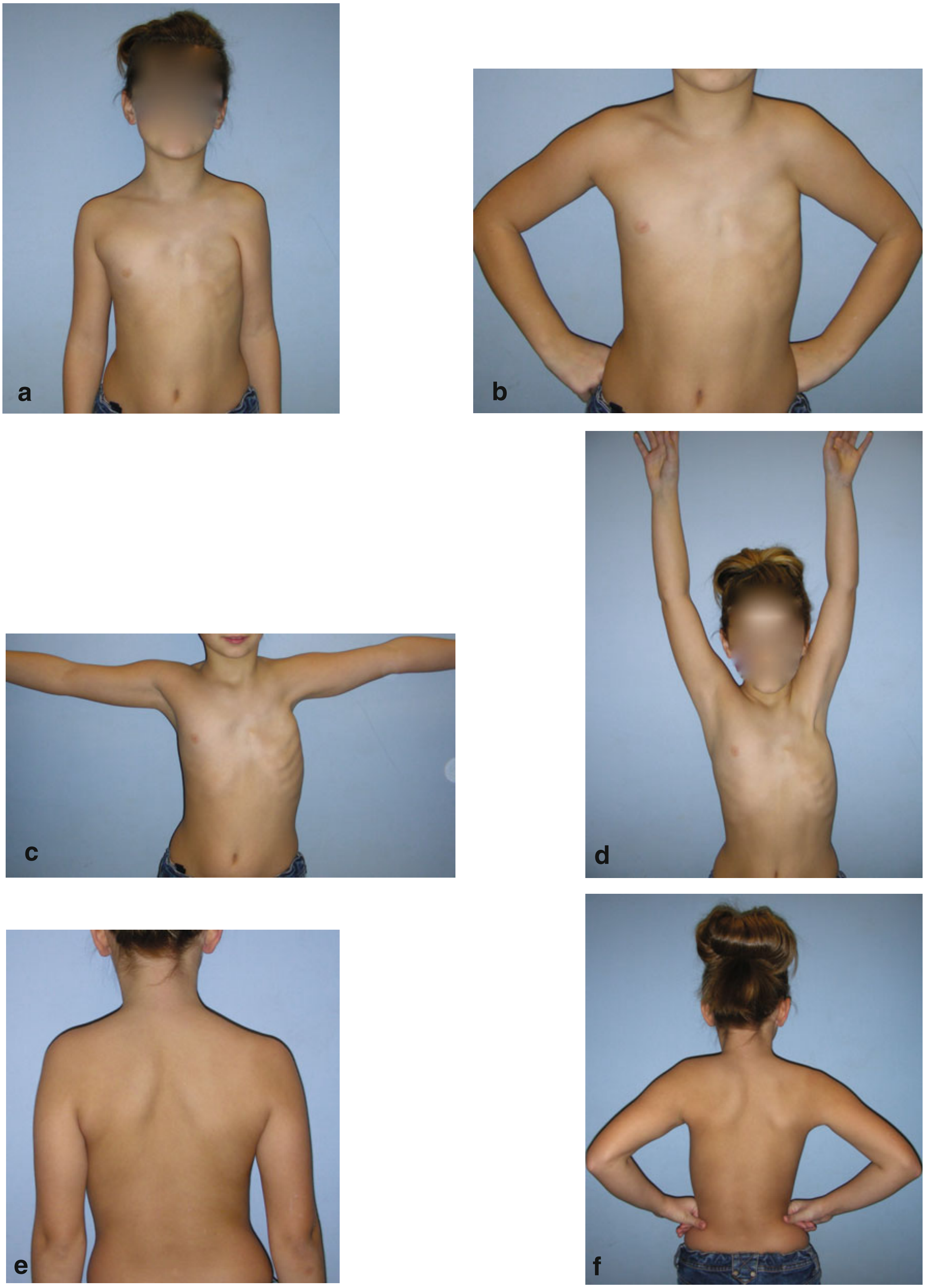


Fig. 13a–f

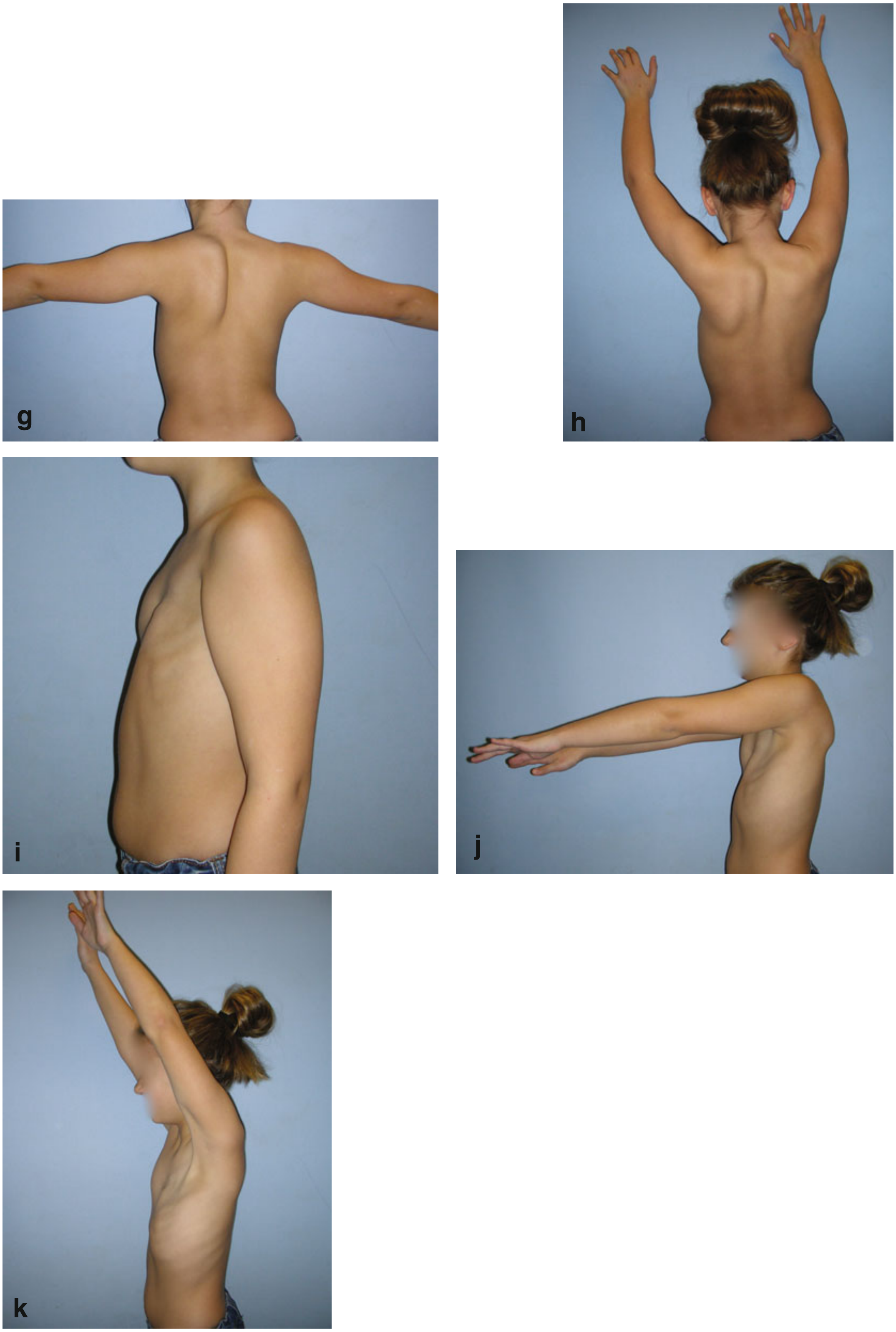


Fig. 13g–k

3.2 Thorax-caliper

Anton H. Schwabegger

Until now, for the documentation of results of the morphologic variations after thoracoplasties photography as a standard measure still remains unsurpassed. Unfortunately, therewith only a planar, two-dimensional image and documentation succeeds, even if pictures are taken in different planes and angles (Chapter 3.1). A far better representation and documentation of the variations of the intra- and extrathoracal diameter in a certain plain, above all in the funnel chest deformity, succeed with computer tomography and therefrom with calculation of the pectus severity index (PSI). This, also as Haller index (Chapter 3.4) well-known reading [6] corresponds only to the internal diameters of the thoracic cavity and serves for the evaluation if a thoracoplasty caused its expansion. On the other hand, this index in certain institutions also primarily substantiates the indication setting for the surgical correction of the funnel chest deformity with remodeling procedures for medical reasons, above all, if restrictions of cardial or pulmonary functions exist. The PSI therefore predominantly serves for preoperative diagnostics and documentation in the funnel chest deformity but hardly in its postoperative course observation and documentation. Furthermore, it finds no place in the diagnostics, indication setting, and follow-up documentation in all other thoracic wall deformities, like Poland's syndrome or keel chest deformity. Using the method of the surface scanning, represented in Chapter 13, the outstanding possibility of an exact 3D-depiction and verification of the thoracic wall morphology, and ensuing postoperative results by calculation of variations of volumes exist indeed [4, 8, 10]. However, disadvantageously an altered nourishment condition may interfere in a false positive or negative extent, causing incorrect optically measured result. Moreover this technology still remains relatively cost-intensive until now.

A further anthropometric method measuring body surfaces and postoperative variations, used in funnel chest patients, is the pectus excavatum anthropometric index [2] (PEX-AI), developed at the Department of Thoracic Surgery of the Heart Institute, Sao Paulo School of Medicine. In order to calculate this index, one requires two indicators. The indicator A on the one hand describes the largest antero-posterior distance; and the indicator B on the other hand measures the innermost point of the skin surface of the pectus excavatum deformity. The measurements are determined in supine position on a flat table and during deep inspiration. In patients with a PEX-AI below 0.12 no deformity subsists and can be estimated as naturally shaped, whereas in contrast to patients with a PEX-AI above 0.12 a pectus excavatum deformity may be demonstrated. Also this method of measurement requires technically expensive devices and relatively much time.

Furthermore Chang et al. [3] in 2009 described another noninvasive anthropometric tool of pre- and postoperative measurement using thermal plastic strips. Using that molding, they were able to follow up the postoperative course even after removal of the pectus bar and could show a significant decrease of the surgically achieved modeling of the anterior thoracic wall in pectus excavatum deformity, after removal of the metal

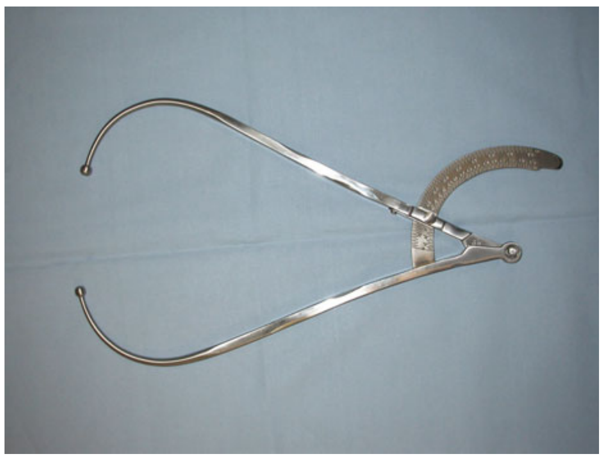


Fig. 1. Thorax-caliper made of steel, with a scale gauged in inches and centimeters. For validity of measurements the spheric ends of the branches must be moderately pressed against skeletal subcutaneous structures

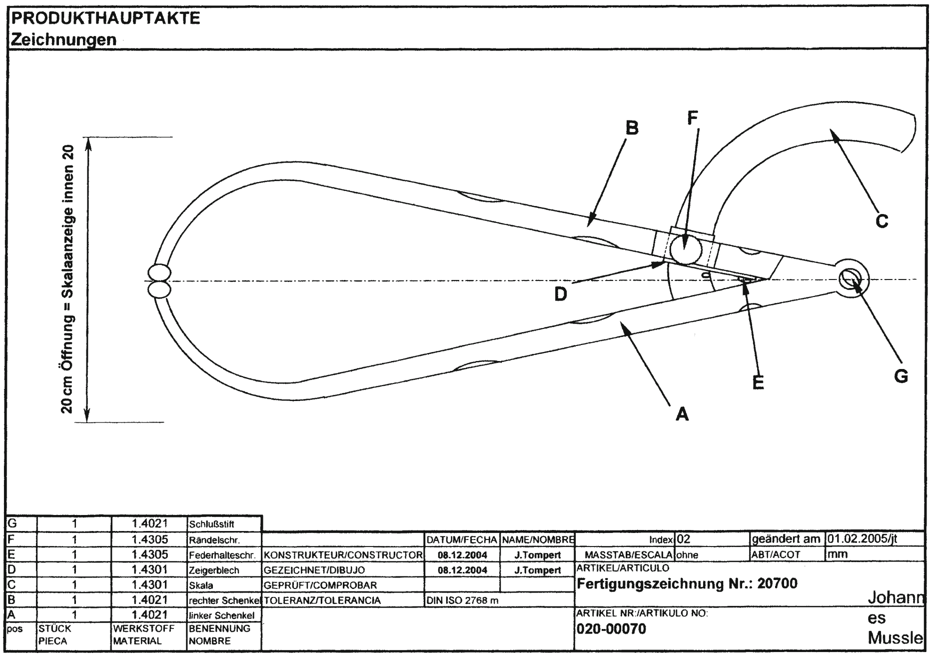


Fig. 2. Construction outline of the thorax-caliper

strut. They found this tool very helpful in further planning of the exact site of pectus bar implantation.

However, alternatively to these techniques another noninvasive method of measuring external diameters is available. It represents a very economical, effective method, meaning only slightest expense, equivalent and in some aspects even superior to the above-mentioned methods in documenting a situs or the course of morphologic variations objectively. It concerns a modification of the pelvis-caliper commonly used in obstetrics and mentioned for the use in pectus excavatum surgery by Haller in 1978 [5]. Ebstein successfully used it already in 1882 [7] for the measurement of thoracic diameters. This above-mentioned pelvis-caliper nowadays was adapted and newly constructed for the application at the thorax (Fig. 1). It is named thorax-caliper and distributed by the company MedXpertTM (MedXpert GmbH, Heitersheim, Germany), which developed this device of avail (Fig. 2).

The basic idea of the development of this thoracic caliper was to create a simple and easily applicable device, which on the one hand circumvents X-ray radiation given by the CT investigation technology, and on the other hand is consistently usable in the clinical daily routine. For the surgeon or the investigator as well as for the patient, this method represents both an objective follow-up observation and an excellent tool for documentation of the outcome quality of shape remodeling. This method of investigation is independent from the person of examiner and the results are always reproducible, also the logistical efforts to organize cost-intensive CT investigations can be circumvented. At times of ever increasing costs in health care, such a simple, cheap, and efficient method for the measurement of thoracic wall deformities can be only of advantage.

Very narrow sagittal diameters on the other hand, previously documented with the thoracic caliper, can yield evidence that the intrathoracic diameter already leads to displacement of the intrathoracal organs and therewith further investigations like computer tomography are indicated necessarily.

3.2.1 Technique of measurement

During measurement, the patient stands upright, with arms hanging downward and remains in loose expirium. The measurement points at the thoracic wall for the transversal diameter are situated at the lateral thoracic wall, at the level of the nipples, exactly between the anterior and posterior axillary line (Fig. 3). Herewith the metallic spheres of the caliper branches are attached to the ribs with moderate pressure in order to grasp fewer soft parts however more the skeletal diameter structures and diameters. The measurement points for the more important sagittal diameters are also situated at the level of the nipples. At the back and at the same level a branch

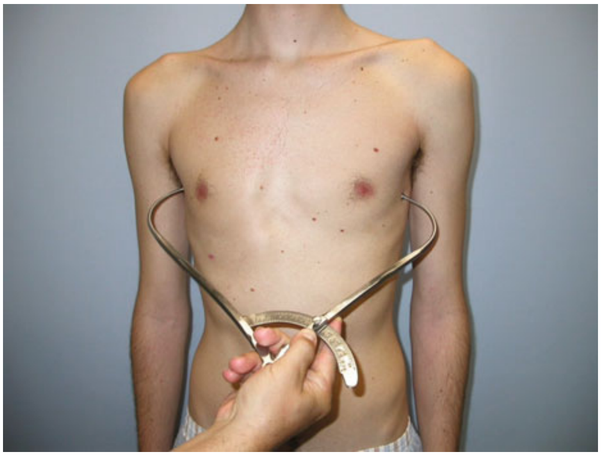
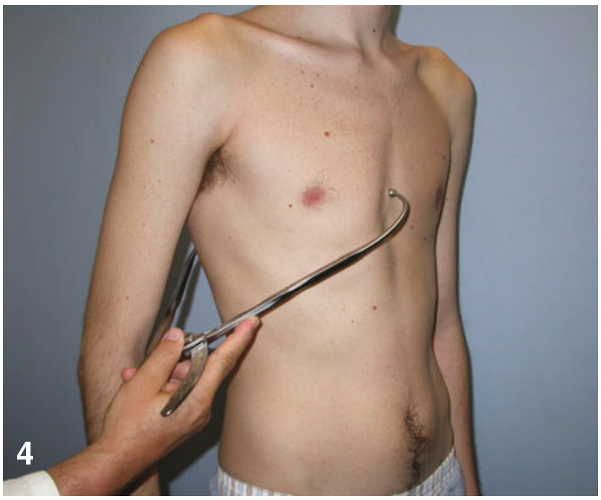
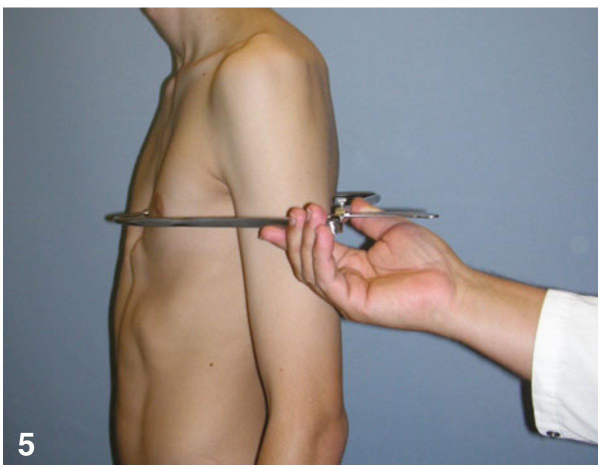


Fig. 3. Transversal measurement taken at a horizontal level of the nipples between the anterior and posterior axillary line

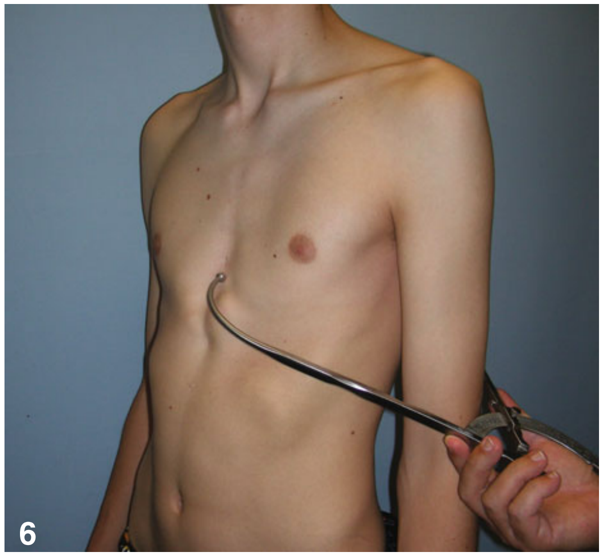


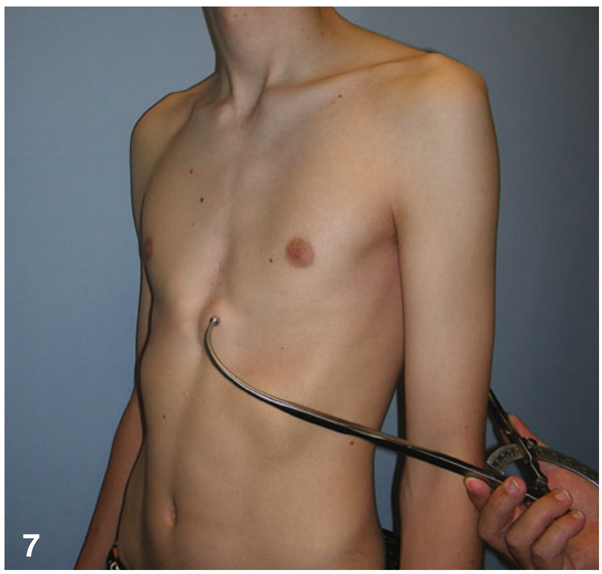


Figs. 4 and 5. Sagittal measurement taken at the same level as the transversal measurement

of the caliper is put on a spinal process (Figs. 4–6). The choice of the anterior measurement point depends on which deformity exists, moreover also on the sex of the patient. The diameters obtained with the thorax-caliper can be read directly on a gauged scale fixed at the caliper in centimeters and inches (Fig. 1).

In male patients with symmetric or only slightly asymmetric deformities the second (anterior) caliper branch to scale the sagittal diameter is placed at the level of the nipples, exactly in a median line above the sternum. If asymmetry or a more rare atypical





Figs. 6 and 7. In cases with irregular surface of the deformity, measurements should be taken at the deepest depression and at the highest point

deformity is existent, setting of additional measurement points is recommended, e.g., at the deepest depression (Fig. 6) and highest elevations of the sternum or ribs deformity (Fig. 7) whereby the measurements must always be taken in a standardized manner in a horizontal plane.
In patients with ptotic or asymmetric breast, the sagittal measurement cannot be carried out at the level of sagged or asymmetrically positioned nipples. The highest point of a keel and the deepest point of a funnel deformity then must be measured and documented, respectively, irrelevant from the level of the nipples, however, in a horizontal plane anyway. By means of photographic documentation, which is carried out in every patient, the location of measurement points can be reviewed retrospectively, where they were taken preoperatively. In order to include the influence of growth and weight alteration on the variations of the chest in youthful patients, also these variations are documented by means of the parameters size, weight,

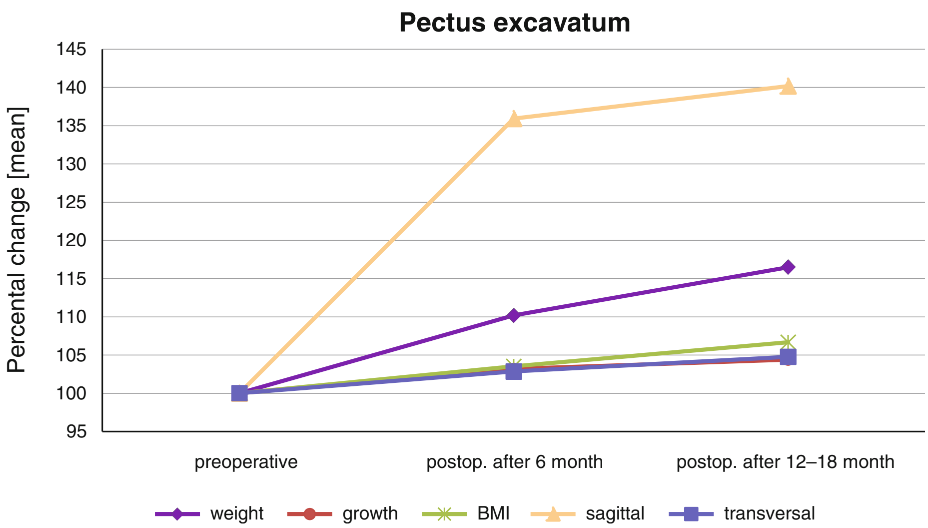


Fig. 8. Measurements result of alterations of the sagittal and transversal diameters in 24 patients with funnel chest deformity from preoperative situs to long-term postoperative follow-up, set in relation to change in weight and height

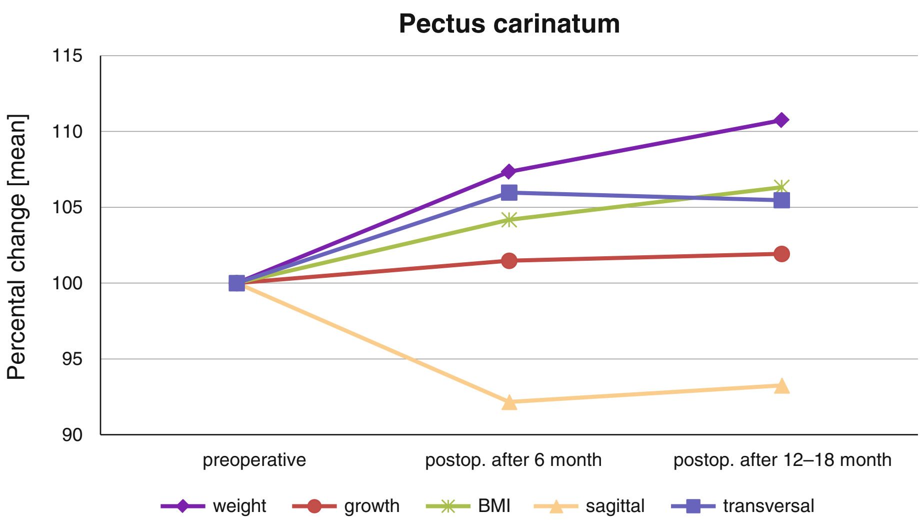


Fig. 9. Measurements result of alterations of the sagittal and transversal diameters in 23 patients with keel chest deformity from preoperative situs to long-term postoperative follow-up, set in relation to change in weight and height

and BMI over the defined period of follow-up. Therefore variations in proportion during growth can be compared and can be brought in reference to the indexes of the changing thoracic diameters (Figs. 8 and 9) statistics.

In a retrospective course study prepared with statistical methods, the thorax-caliper has proved itself as extremely useful to exactly compare the course of preoperative to immediate postoperative situations as well as to document the long-term course [1]. The measurement with the thorax-caliper proved itself substantially superior compared to the remaining measurement methods, because it allows a close-meshed, anytime repeatable, very economical, easily learnable, painless, and above all noninvasive survey. The thorax-caliper though cannot replace a preoperative imaging of the intrathoracal situs necessary in certain cases, yet the indications for these investigation techniques are anyway different. The CT investigation on the other hand should not be requisitioned for the pure course observation because of the radiation burden and its costs.

For the patient himself, a rapid and easily available course observation of the postoperative result ensues, whose positive subjective estimation through the patient substantially depends on visibility and objectifiable results. Through the straightforward measurement with the thorax-caliper, these results are easily understandable for the patient and in this manner perhaps contribute to promotion of the recovery process, within which the psychic component may not be disregarded. Positive messages, in the sense of an improvement of the measurable values, obtainable through the thorax-caliper will allow an essential influence on the psychic contentment and the quality of life of the affected patients for certain [9] (Chapter 3.6).

Especially for the patient, the investigation itself works simply and is feasible without any preparation. For the investigator, under the prerequisite that he complies with the given measurement criteria, no special education or sophisticated instruction is necessary.

3.2.2 Conclusion

Comprehensively the measurement with the thorax-caliper serves less to the indication setting for surgery, but rather enables a straightforward documentation and course observation as well as it represents a supplement to photographic documentation in the sense of quality control and quality securing of such remodeling surgical interventions.

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3.3 Radiologic diagnostics

Michael Rieger

Pathologic processes that may involve the chest wall include congenital and developmental anomalies, inflammatory and infectious diseases, and soft-tissue or bone tumors. Many of theses processes have characteristic radiologic appearances that allow already definitive diagnosis. Sternal deformities on the one hand can be visualized with conventional radiography alone, on the other hand their severity may be quantified with computed tomography (CT).

3.3.1 Conventional radiographs

In funnel chest (pectus excavatum), the most common congenital thoracic deformity, the sternum is depressed so that the ribs on each side seem to protrude more anteriorly than the sternum itself. This sternal and cartilaginous depression reduces the prevertebral space, resulting in leftward displacement and axial rotation of the heart-simulating cardiomegaly, which can be depicted in the posteroanterior chest radiography (Fig. 1), as well as reduction of the space occupied by the left lung. The displaced pulmonary vascularity and obscured right cardiac border resemble a right middle lobe process. The posterior ribs run horizontally, whereas the anterior ribs show a more oblique course downward. The degree of a sternal depression is easily visualized at lateral radiography (Fig. 2), but not in the posteroanterior view.

Keel chest (pectus carinatum) is a deformity of the chest wall in which the sternum exhibits abnormal protrusion anteriorly (Fig. 3). A relatively large sagittal diameter is observed on lateral radiographs. On frontal radiographs, the lateral portion of each rib appears elongated and straight downward [1, 2].

Funnel and keel chest can be diagnosed on conventional radiographs; however, the lateral radiography clearly depicts the reduced or enlarged sagittal diameter in these congenital deformities, whereas the posteroanterior chest radiography shows only indirect signs of a present skeletal deformity, like displaced heart or lung anomaly. The degree and characterization of the deformity, the relationship of the sternum to the bony part of the ribs, and their cartilages as well as the deviation of these structures in various planes thus may be much better depicted with CT.

3.3.2 CT-scan

The rapid development of helical CT has resulted in exciting new applications for CT. The technical enhancements that multi-detector row computed tomography (MDCT) devices have experienced over the past years have brought significant technical progress and

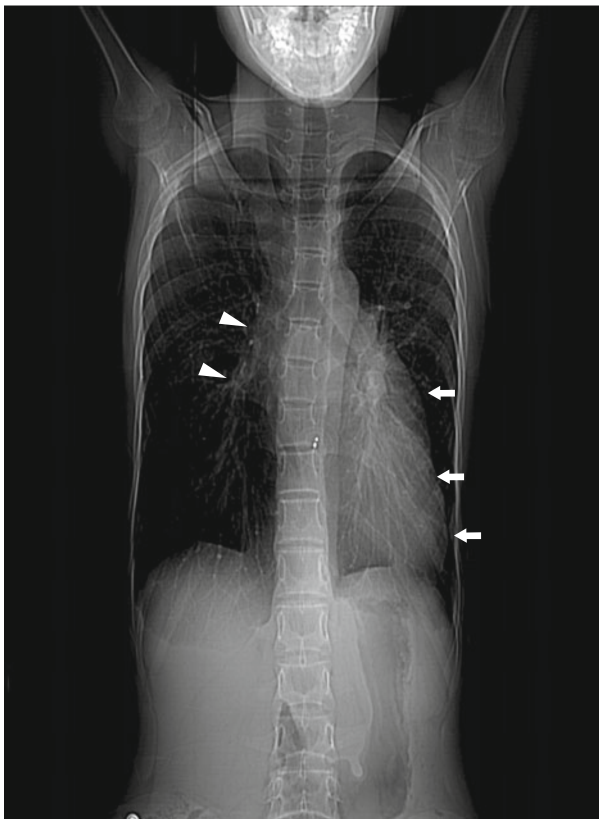

Fig. 1. Pectus excavatum in a 14-year-old girl. Coronal CT scout of the chest shows leftward displacement of the heart (arrows). Because of the rotation of the heart the pulmonary arteries of the right side are depicted prominently (arrowheads)

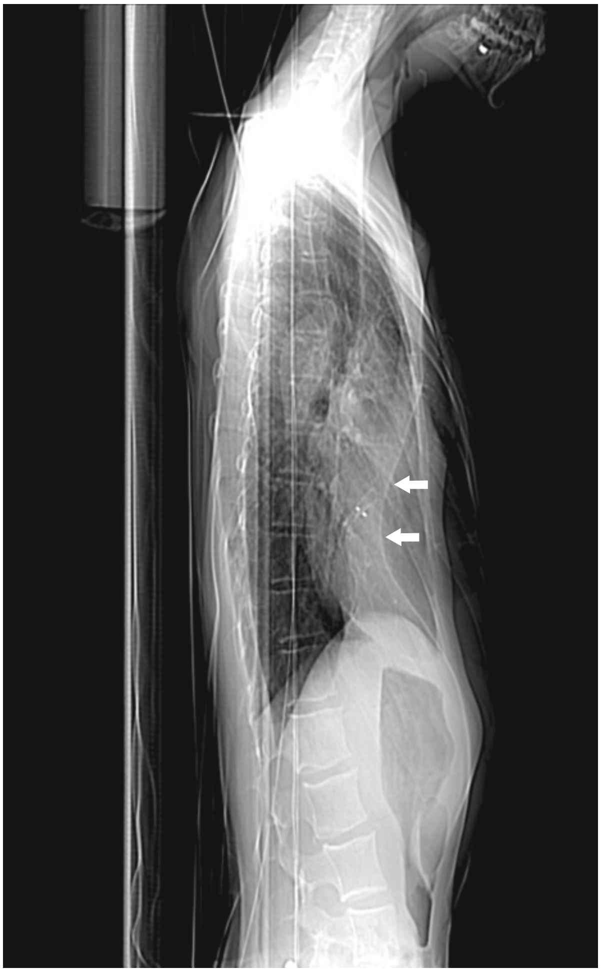

Fig. 2. Pectus excavatum in a 14-year-old girl (same patient as in Fig. 1). Sagittal CT scout of the chest demonstrates severe depression of the sternum (arrows)

Fig. 3. Pectus carinatum (keel chest) deformity. An extended sagittal diameter is present, with protrusion of the sternum

influence in all fields of diagnostic radiology. Equipped with multiple row detector arrays resulting in increased longitudinal volume coverage and thin collimation, MDCT provides a volumetric scan of the entire thorax in only one short breath hold (less than 10 s). This provides true isotropic as small as 0.35 mm resolution in all directions with reduced artifacts due to cardiac and respiratory motions. With volumetric data acquisition, it is now possible to reconstruct contiguous thin-section images as well as volume rendered and multiplanar reformatted images. Using a 64-row MDCT for example the quality of coronal and sagittal reformations is comparable to axial images. Coronal reformations on 64-row MDCT show substantial agreement with axial images in the evaluation of thoracic abnormalities [3].

Because of the potential for malignancy, children who have lumps palpated in the chest wall at physical examination are often referred for cross-sectional imaging. It has been shown that in most of these children, the cause of the lump is a benign variation in the anatomy of the anterior chest wall, which has a number of functions that when altered can result in a variety of pathologic processes. Because the chest wall provides a structural framework that is the basis for physiologic motion of the pulmonary system, deformity of the chest wall can result in both restrictive and obstructive lung diseases. The space in the superior mediastinum normally is limited, confined by the sternum superiorly and the vertebral column posteriorly. Abnormal thoracic configuration can result in further reduction in the volume of this space or in alteration in the anatomic relationship between the trachea and adjacent structures. Both mechanisms can result in displacement and compression of the mediastinal structures (Fig. 4). The most common abnormalities of chest wall configuration result from overgrowth of the length of costal cartilages which occurs in utero and does not allow the sternum to come forward with inhalation [4] (Fig. 5). Although

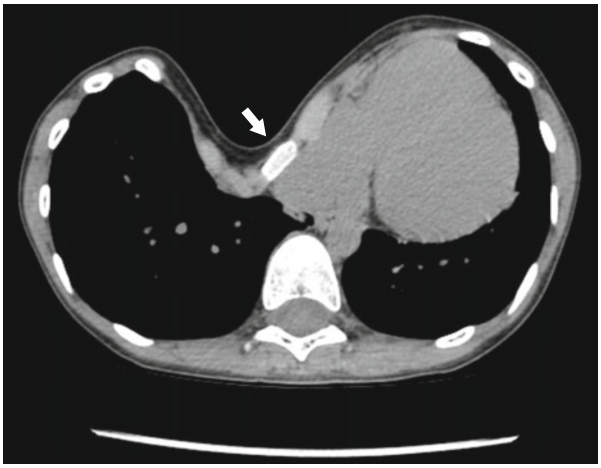

Fig. 4. Pectus excavatum in a 14-year-old girl (same patient as in Fig. 1). Axial CT scan shows anterior chest wall to be concave. The midline chest wall (arrow) is located much more posteriorly than the more lateral chest wall. The heart is hard-pressed and toward left lateral displaced

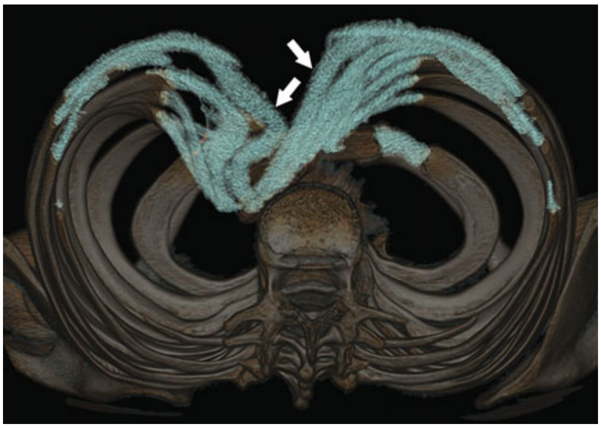

Fig. 5. Pectus excavatum in a 14-year-old girl (same patient as in Fig. 1). Oblique transversal 3D volume rendering reconstruction depicts the deformity of the chest wall with depression of the sternum and especially from an overgrowth of the length of costal cartilages (arrows)

Fig. 6. Pectus excavatum in a 14-year-old girl (same patient as in Fig. 1). 3D volume rendering reconstruction with a low threshold showing the patient's skin surface depicts the cosmetic problem in this patient due to this deformity of the chest wall

most problems that are due to these deformities are aesthetic (Fig. 6), pectus excavatum deformities can cause chest pain, fatigability, dyspnea on exertion, palpitations, and restrictive lung disease. Depending on the extent of deformity, various methods of surgical repair to be performed are available [5].

For the purpose of providing information to the surgeons, MDCT allows exact pre-op measurement of the deformed bony and cartilaginous rib cage and thus supports the operative correction and placement of reconstructive maneuvers. A transverse section of the MDCT data may be used exemplarily to compute the "pectus index" advocated by Haller et al. for quantification of the chest wall defect in pectus excavatum [6]. A transverse section at the level of the greatest severity of the defect may be used to determine this ratio of the lateral thoracic diameter to the sagittal diameter as a ratio of more than 3.25 is estimated as pathologic. In addition, MDCT depicts the displacement of mediastinal structures caused by the abnormal thoracic configuration and their consecutive probable impairment. Furthermore, pulmonary changes like areas of emphysema or hypoventilation can be depicted within the same MDCT examination, which offers therefore a comprehensive workup of patients suffering from congenital thoracic wall deformities.

Using a recent CT device and a low dose protocol the effective radiation dose of a CT examination of the chest is about 3.2 mSv with the highest values for the breast (about 8.1 mSv) and nearly no uterus radiation.

3.3.3 Three-dimensional volume rendering CT reconstruction

Nowadays the common volume rendering (VR) technique makes use of the entire data set and thus conveys more detailed information than other 3D rendering techniques. In VR, the contributions of each voxel (volumetric pixel) along a line of sight from the viewer's eye through the data set are summarized. This process is repeated many times to determine each pixel value in the displayed image. VR can show multiple internal and overlying features, and the displayed intensity is related to the amount of bone encountered along extending through the volume. The flexibility of the VR algorithm allows the radiologist to tailor the degree of surface shading and bony opacity to the actual clinical problem [7]. In patients suffering from congenital thoracic wall deformities, the VR reconstruction mode allows far better than all other reconstruction methods depiction not only of the osseous structures of the thoracic cage but also of the particular structures of interest for the remodeling surgeon, namely the rib cartilages and if necessary of other thoracic structures. In a patient who is referred for cross-sectional imaging, unenhanced helical 64-detector row MDCT (LightSpeed VCT, GE, Milwaukee) is performed. Typical parameters include a slice thickness of 0.625 mm, volume coverage of 40 mm/rotation with a minimum rotation-time of 0.35 s, and a weight-based low tube current technique. Using the integrated transfer rates with gigabit connectivity, the large data volume generated by routine sub-millimeter (0.625 mm) scanning is automatically transferred within a few seconds together with automatically created multiplanar reformatted images ("direct MPRs") to a 3D rendering workstation (HP Workstation xw 8000) running Advantage Windows software (Advantage Windows 4.2, GE Healthcare). The ability to directly create MPRs from the source data allows radiologists to visualize CT images online in three planes. 3D VR reconstructions are promptly managed using stored presets. After image generation, the whole data set is sent to a "Picture Archiving and Communication System" (PACS, J-Vision, TIANI, Austria).

Often, the presence of an anatomic variation will be subtle on standard axial CT images alone. This finding is particularly true of prominent convexity of the anterior rib or costal cartilage, prominent asymmetric costal cartilage, tilted sternum and its associated findings, and other rib anomalies. It may be difficult on axial images alone to show that the anomaly at the level of the palpable lump is more anterior than the ribs and the costal cartilages superior and inferior to that level. 3D reconstructions of the helical CT images are helpful to identify and to define such lesions (Fig. 5). Volume rendered images with a low threshold showing the patient's skin surface can be used to identify the protrusion palpated on physical examination (Fig. 6). By increasing the threshold and concomitantly semitransparent depiction of the skin, the underlying bone and cartilage can be demonstrated and be used to correlate the anatomic variation with the exact site of the palpable lump (Figs. 7–9). In some cases, several anatomic anomalies lead to the finding of a palpable protrusion. For example, a tilted sternum may be associated with a subluxed clavicular head or costal cartilage, or a rib anomaly can be associated with the adjacent rib protruding anteriorly and cause a palpable lesion. These combined anomalies may involve several levels of ribs or costal cartilage and are often more easily seen on 3D images. 3D images are also helpful if the palpated lesion is caused by a disease process. The relationship between the mass and the adjacent ribs can be further depicted. Transversal MDCT images together with 3D VR reconstructions can define the severity of the deformity and

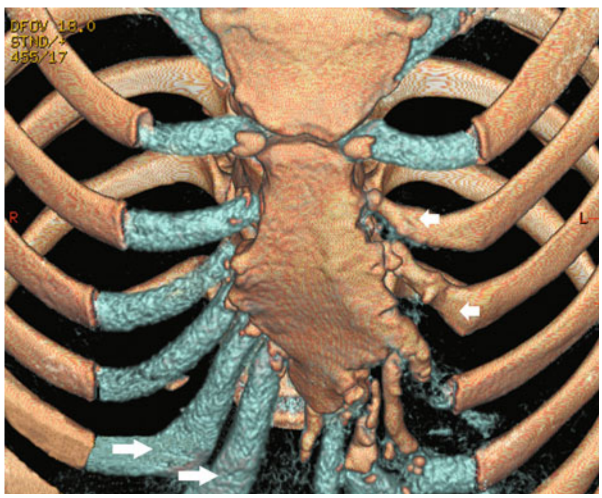

Fig. 7. Pectus excavatum in a 33-year-old woman. Oblique coronal 3D volume rendering reconstruction depicts the overgrowth of the length of the lower costal cartilages (arrows), which causes the deformity of the sternum with a sunken xyphoid. In addition the cartilages of the third to fifth ribs are missing because of complete ossification (short arrows)

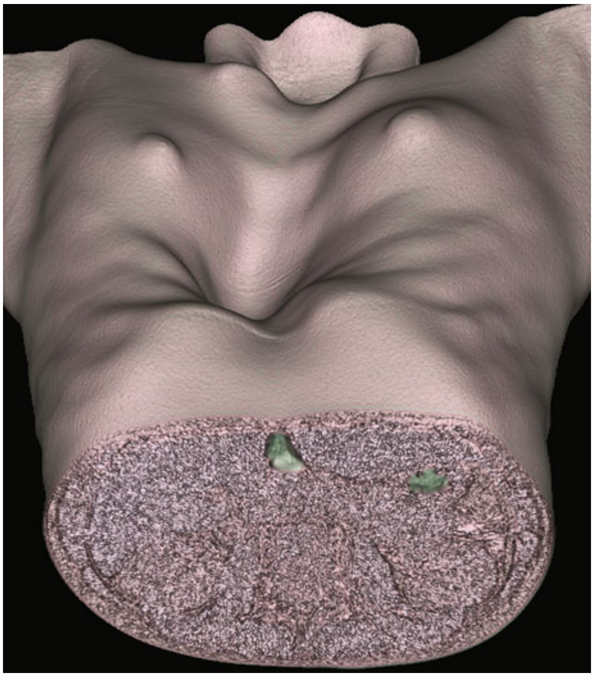

Fig. 8. Pectus excavatum in a 14-year-old girl (same patient as in Fig. 1). Oblique coronal 3D volume rendering reconstruction with a low threshold showing the patient's skin surface can be used to identify the protrusion palpated on physical examination

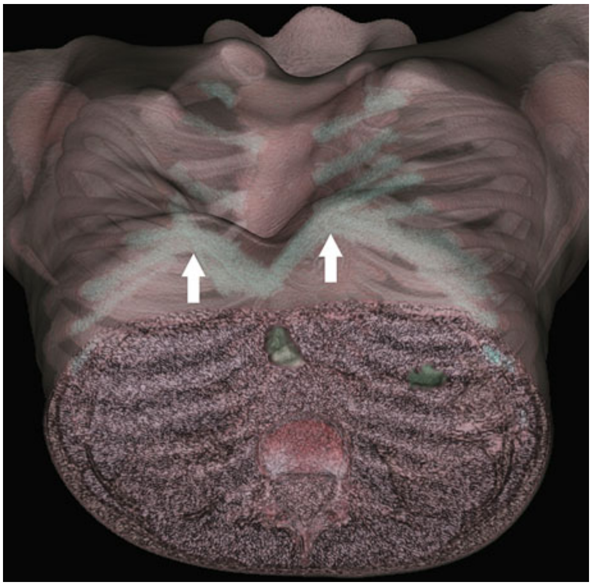

Fig. 9. Pectus excavatum in a 14-year-old girl (same patient as in Fig. 1). Oblique coronal 3D volume rendering reconstruction with a slightly increased threshold and concomitantly semitransparent depiction of the skin demonstrates the underlying bone and cartilage and can be used to correlate the anatomic variation with the exact site of the palpable lump (arrows)

identify any anomalously positioned cardiovascular structures that may complicate implantation of a pectus repair device.

Evaluation of funnel and keel chest is also possible using magnetic resonance (MR). This modality depicts sternum, ribs and cartilage, a one-stop shopping similar to CT, which includes the evaluation of all structures of the thorax, is not possible. The advantage of MR over CT and radiographs is an examination without radiation. Performance of 2D reformations in different planes is possible, 3D reconstructions like VR are not available. The image acquisition time for a chest MR (15–20 min) is much longer than a CT examination and therefore not accomplishable in one breath hold.

In conclusion, 3D reconstructed helical MDCT images assist in the evaluation of children with potential chest wall abnormalities or anatomic variation. Because of the orientation of many of these variations, the information displayed on 3D images may be helpful in identification of the anatomic variation, in communication of the presence of these benign anomalies to referring physicians, and in the development of understanding of these anomalies [8].

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3.4 Haller index, pectus-severity-index

Anton H. Schwabegger

A so-called fronto-sagittal-index and a vertebral index were described by Backer [1] already in 1961, using conventional radiographs for the measurement and assessment of preoperative situs compared with post-operative results. Hümmer in 1972 [5] described a so-called "Trichterbrust-Index" meaning a funnel chest index, calculated as a qoutient of sagittal radiographic diameters of the cranial manubrium ridge to the spinal process and the deepest depression to the spinal process. Such a similar index is described and refined nowadays, known as the Haller index, however using other techniques and measurement planes. This, in other words named pectus-severity-index (PSI), is calculated from an axial computed tomography (CT)-scan [3] or MRI [4]. It means the ratio (Fig. 1) between the largest transversal (internal) thoracic diameter and the shortest (sagittal) internal vertebro-sternal distance (*a*) at the position of the deepest inclination of the sternum (*b*) (PSI = a/b). In asymmetric cases, when ribs bulge inward parasternally and represent the deepest point at the thoracic wall, the internal diameter is measured from this point to a tangential horizontal line (Fig. 2) from the subjacent vertebral body [3].
A criterion of Nuss [6] lies therein, that an index with the value of 3.2 or more represents the cutoff, which defines a funnel chest deformity as moderate or severe. Values below that on the other hand yet mean no indication for invasive or mininmal invasive remodeling surgery to correct such a deformity. Daunt et al. [2] however estimates the value of 2.7 or higher, which determines a necessity of surgical treatment of the pectus excavatum deformity.
Moreover, the Haller index is dependent on age and sex of the patients and shows large variability in relation to the entire body anatomy [2, 7]. Furthermore, the anatomy of the thorax concerning its depth and width changes significantly with growth from childhood over puberty to adolescence and therefore represents a dynamic parameter. Especially during puberty, growth spurts lead to altered anatomic measures, at a time, in which, at least in our collection of patients, predominantly seek for and desire correction of that annoying stigma. Therefore within that time-frame only limited usefulness exists to apply the Haller index as a means for selection of an indication for or against any surgical intevention.
In any case, the index can serve as an additional decision-making aid to clinical diagnostics, when

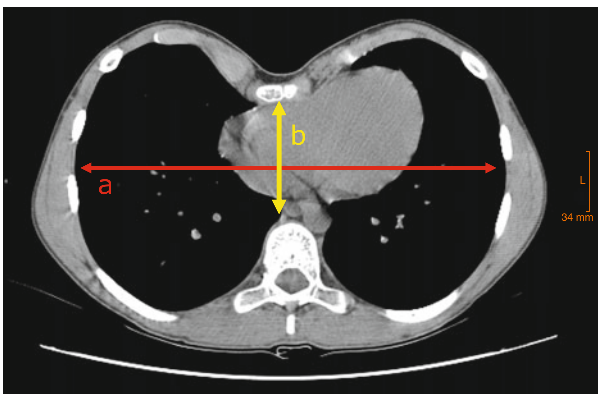

Fig. 1. Haller index in a symmetric case. *a* means the maximal horizontal exact transversal diameter and *b* is the minimal sagittal diameter, measured from the sternum to the subjacent vertebral body

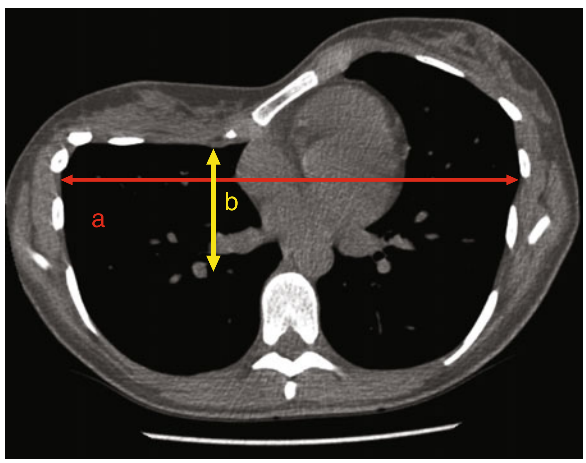

Fig. 2. In asymmetric cases *a* is measured as depicted in Fig. 1, wheras *b* is measured from the deepest point of deformity to a horizontal tangential line from the adjacent vertebral body

Fig. 3. "Dextrocardia" in a female patient with Noonan syndrome and moderate funnel chest deformity. Shifting of the heart and greater vessels to the right side, eventually due to the depression of the sternum

other investigations such as ergometry, spirometry, and psychologic diagnostics allow no conclusive decision for or against a surgical intervention. CT is helpful only in these patients, in whom thoracoplasty is required for expansion of the thoracic cage. The radiologic investigation is necessary in the more severe cases of deformity anyway, to depict the relationship of the intrathoracal organs to the skeletal structures exactly. A major depression of the sternum causes a shifting of the organs, usually further to the left, but can also lead to transposition to the right side, for example in situs inversus (dextrocardia) (Fig. 3). Particularly in cases with additional deformities at the trunk or in syndromal cases, CT figures are definitely necessary for an exact operation plan anyway to avoid injuries of intrathoracal structures when anatomical variations might be present.

The advantages of CT slices lie herein that plains of special interest can be read to determine the PSI very exactly and to exclude endangering anatomic variations. The disadvantages on the other hand remain therein that for postoperative follow-up inspections, further CT investigations were necessary, with the problematic nature of organizational expense and emerging costs of health care as well as the additional burden of radiation.

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3.5 Cardiopulmonal investigation

Barbara Semenitz

In patients with marked funnel chest deformity frequently impairment of fitness exists that can be objectified by means of a symptom-limited ergometry [8–10]. The most frequent causes for the fitness impairment are the thoracic wall deformity itself or a training deficiency and/or the combination of both. Moreover, citations are found in the literature that in 3% of patients with funnel chest deformity also a congenital cardial vitium coexists. The extent of fitness restriction in these particular cases with a cardial vitium has to be investigated in an individual and more distinct process apart from the standard measurements for the collection of patients with funnel chest deformity [3]. However, training deficiency is often also the consequence of a motivation problem because many of these patients perceive their thoracic deformity as a massive physical deficit and above all for psychologic reasons avoid participating in almost all kinds of sporting activities.

The functional restriction of fitness through the thoracic wall deformity explains itself by the reduction of distance between the sternum and the vertebral column and hence negative hemodynamic effects follow [1, 2, 4]. During physical exercise and increasing breathing movements of the thoracic cage, a sunken sternum is not capable of following the respiratory thoracic wall excursions and thus adversely restricts inflation of the lungs and ensuing cardiac output. During forced inspiration and contraction of the diaphragm, even retraction of the sternum and ensuing paradoxical breathing may be caused [6]. At the same time, based on the distortion of the heart, an impairment of the venous backflow from the Vena cava inferior into the right atrium and ventricle occurs. This again decreases the heart minute volume and therewith the capability of adaptation of the cardiac output with increasing physical exercise [10]. This impairment is observed frequently, and the distance between sternum and vertebral column is lesser than 10 cm. Below a distance of 8 cm, this restriction of cardiac output power is almost always present [10].

A further mechanical problem results from the increased strenuous effort, which is necessary to lift the sternum and to create therewith the prerequisite for cardiopulmonal adaptation to an increased demand of respiration during physical exercise [6, 11]. From this viewpoint, a measurement scale for the extent of functional impairment in a funnel chest deformity is available with the pectus-severity-index [5, 7]. The pectus-severity-index (Chapter 3.4) is calculated from an axial CT scan. It is the ratio between the largest transversal (internal) thoracic diameter and the shortest (sagittal) vertebro-sternal distance (a) at the position of the deepest inclination of the sternum (b) (PSI $= a/b$).

Also the position of the torso during investigation (ergometry) very substantially influences these sketched mechanisms of fitness alterations. In the case of a healthy patient, sufficient compensation mechanisms exist to largely equalize the efficiency in the lying (horizontal) and the sitting position during ergometric stress. However even in these healthy probationers in the lying position, the performance always is lesser, but rather modestly [3, 10]. In patients with a funnel chest deformity on the other hand, negative effects on hemodynamics are clearly more marked in the sitting than in the lying position. Ensuing from that and as Liese and Bühlmann [10] report, the heartbeat volume during the sitting examination is 20% lower than in the lying position of examination in patients with a marked funnel chest deformity.

In their investigations Liese and Bühlmann were able to differentiate between training deficiency and fitness impairment through the funnel chest deformity utilizing a submaximal performance test. At a heart frequency of 170/min either the effective working capacity (EWC) or the physical working capacity (PWC) 170 is determined by means of a stepwise increasing test on the bicycle-ergometer, first sitting, and subsequently in a lying position [10].

The comparison of the nominal output to be expected in Watt, which represents a submaximal stress, with the actual wattage permits the judgment of the effective

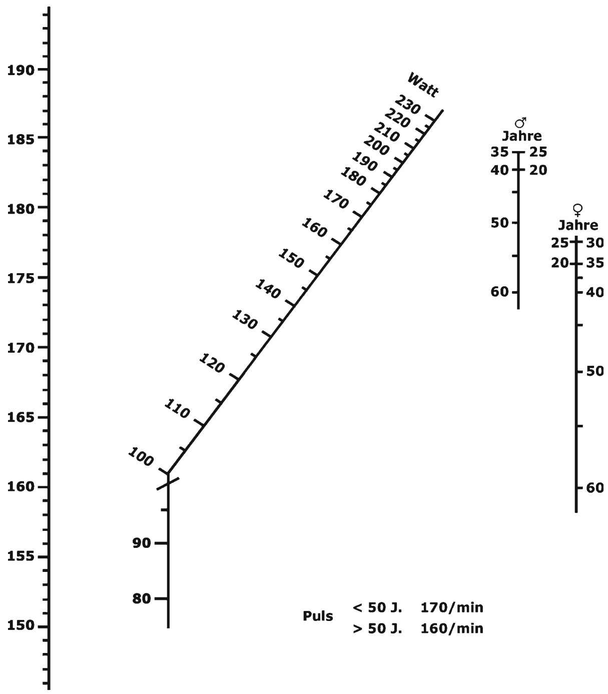


Fig. 1. Nomogram

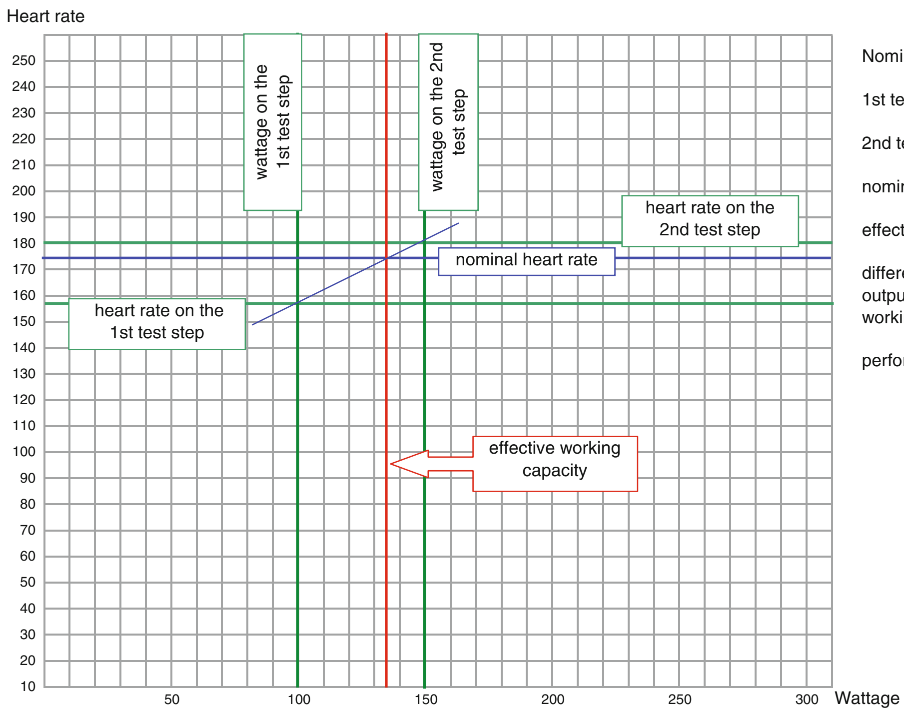
Table 1: Age-dependent calculated heart rate

Age	Heart rate	Age	Heart rate
15	187	45	161
16	186	46	161
17	185	47	160
18	185	48	159
19	184	49	158
20	183	50	157
21	182	51	156
22	181	52	155
23	180	53	155
24	179	54	154
25	179	55	153
26	178	56	152
27	177	57	151
28	176	58	150
29	175	59	149
30	174	60	149
31	173	61	148
32	173	62	147
33	172	63	146
34	171	64	145
35	170	65	144
36	169	66	143
37	168	67	142
38	167	68	142
39	167	69	141
40	166	70	140
41	165	71	139
42	164	72	138
43	163	73	137
44	162	74	136

work capacity. The nominal output is dependent on age, height, and gender. Subsequently the nominal heart beat rate according to the formula: $170 - 0.86 \times (\text{age in years} - 35)$ is calculated or taken out from a corresponding table (Fig. 1).

The physical stress is carried out as a two-step test, whereby the load of the first step should remain 25 W below the nominal wattage and that of the second step 25 W above the nominal wattage, so that the difference of the steps counts 50 W. The duration of stress per step lasts five, but at least 3 min [10]. Our practical experiences however showed that nominal wattage calculated or derived from Table 1 very often already exceeds the actual maximal performance. Therefore a determination of maximal performance by means of bicycle-ergometry of each individual patient prior to a submaximal standardized test seems to be ideal, to enable determination of the individual wattage for the submaximal stress test then. The maximal and the submaximal stress tests in the sitting and lying position would have to take place however at different days to exclude an influence of the result of the two-step tests through the maximal stress test. This procedure in many cases, however, is hardly feasible for organizational reasons like distant residency of the patients. We therefore include the case history into an estimation of the physical efficiency of the patient and correspondingly adapt the calculated wattage. For the case that the levels of the two-step test in the sitting position were selected to high and the test therefore is interrupted by the patient before a time limit of 6 min, a repetition of the test with decreased stress levels is required after a recess of at least 2 h, which extends the total test duration correspondingly.

Between that two-step test in the sitting and then in the lying position, a break should be kept for also at least 2 h. During the stress tests itself, heart rate and blood pressure are measured in 1-min intervals. The maximal heart rates of the first and the second test steps as well as



Nominal output expected	228	wattage
1st test step	100	wattage
2nd test step	150	wattage
nominal heart rate	174	wattage
effective working capacity	135	wattage
difference between nominal output expected and effective working capacity	93	wattage
performance	59%	

Fig. 2. Effective work capacity (exemplarily)

the corresponding wattages are recorded in a coordinate system (heart rate and watt), subsequently the points of measurements are interconnected. The effective work capacity then results from the determination of the wattage at the nominal heart rate (Fig. 2 exemplarily). Alternatively to this method, the determination of the PWC 170 is available.

Likewise to the method described above, testing is carried out in two steps also. However, the wattage in the PWC 170 is determined at a heart rate of 170/min. The nominal (standard) values for the PWC 170 in watt is calculated as follows: for men: kg of body weight × 3, for women: kg of body weight × 2.5.

If the investigations show that the effective work capacity and/or the PWC 170 in the sitting position is/are higher than in the lying position, a high probability is present that no hemodynamic impairment exists based on the funnel chest deformity.

Does it on the other hand turn out that the measured values of cardiopulmonal capacity in the lying position are equal to or higher than in the sitting position, then a functional impairment through the sternal depression is ascertained. The explanation for that effect is, that in the lying function the heart slips more crucially thus away from the sunken sternum, which then results is better hemodynamics and elevated work capacity. This means that corrective thoracoplasty with elevation and stabilizing of the sternum and parasternal ribs will lead to an improvement of hemodynamics and therewith efficiency or fitness.

Despite normal physical efficiency in the sitting position, an impairment of the hemodynamics in the lying position may not be excluded, and that is why we recommend that two-step test both in the sitting and in the lying position. The point of origin of that differentiation in the sitting and then in the lying position is that at that time Bühlmann and Liese recommended the procedure only with patients, whose efficiency was below average. However, actually there exist patients with funnel chest deformity who show a normal physical efficiency in an achievement test, but better in the lying than in the sitting position. If relying only on absolute values of efficiency, patients with hemodynamic impairment cannot be detected by that simple but not comparative investigation.

3.5.1 Conclusion

Differential ergometry testing should always be added to and included into the preoperative judgment of every case of patients with moderate to severe funnel chest deformity. It represents an objectifying measurement of subjectively reported impaired fitness, caused by the deformity. It helps to separate those cases with merely lack of physical training from those with true cardiopulmonal functional deficits. This differential diagnosis and distinguishing between physically based from psychologically triggered impairment of fitness evidently influences the selection of more or less invasive treatment, whether as an aesthetic indication or for functional reasons. In some cases despite apparent funnel depression measured normal fitness and without any signs of hemodynamic impairment may even substantiate the decision not to go for surgery but to suggest physical training, without or with psychotherapeutic support.

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3.6 Psychological investigation

Gerhard Rumpold, Martin Lair

A beautiful body according to the entertainment media in our society guarantees success in all areas of life, because physical attraction is often equated with positive social and intrinsic individual values. On the one hand the abstract term of beauty is predetermined socially, however, on the other hand it is also an intraindividual complex emotional and cognitive process. The subjective body perception shows a multidimensional construct which is determined from affective, cognitive and behavioral aspects. Hence, it may be seen not only as a result of the discrepancy between the ideal and real body perception [3], but it is an essential component of the self-conception and therefore, amongst others, can have far-reaching consequences on the quality of life [5]. Many investigations confirm the assumption that the satisfaction with the own body correlates significantly positively with the self-worth. Thereby, women show a higher correlation than men [12]. Particularly with youngsters a clear connection between body satisfaction and self-esteem can be allocated empirically [7, 18]. Verkuyten [19] could show that in youngsters this correlation is more significant than the connection between the self-esteem and other dimensions of the self-perception, like abilities at school or perceived popularity. The body satisfaction that comes along with self-worth also increases the positive expression of other attributes. In an investigation of students it could be shown that the more content they were with their physical appearance, the more attractive, beautiful, efficient and robust, but less worried and stressed when they judged their body, all the more they accepted it and experienced it as integrated in their self-experience [13]. According to Berscheid et al. [1] persons with above-average body satisfaction describe themselves as more charming, assertive, conscientious and intelligent than test persons with subnormal body satisfaction. In contrast to the impacts of a positive body perception, timidity, unsteadiness, depression, hostility and neurotizismus are significantly associated with body displeasedness [9].

3.6.1 Psychosocial effects of a funnel or keel breast deformity

Patients with a funnel or keel breast deformity often report about feelings of embarrassment and sense of shame based on their constitutional "flaw". Parents mostly observe the suffering of their child and subsequently consult medical attendance. Schwabegger et al. [15] report that the degree of suffering in patients with a very distinctive keel breast deformity often is higher than with a funnel breast deformity, because the keel deformity can poorly be concealed by clothes. Researches yielded that in patients with funnel and keel breast deformity and because of this constitutional appearance, the sense of shame can lead to limited self-worth, feeling of inferiority, depressive mood, coyness, social timidity and maladaptive social behavior [14]. In their investigation Einsiedel and Clausner [6] ascertained the following, to some extent very interfering reaction tendencies: anxiety, shame, constraints, failure focusing, stigmatization feelings, enhanced suggestibility, ambivalent behavior, dissimulation and rationalization trends, limited communication ability, inhibition of autonomous trends, unspecific social problems, nervous restlessness, latent aggressiveness and limited performance. Moreover, in these patients they observed a high focusing of attention to their body, a negative appraisal of their appearance, a disturbed perception of their body shape and occasional depressive reactions. All these psychological symptoms were stronger developed in older patients (>11 years) than in younger ones. Einsiedel and coworkers led back this observation on the increasing socialization and ability to self-reflection in the course of growing-up. Therefore the features of the deformity are not authoritatively determining the individual coping with the disease, but the available psychosocial protective attributes or the individual risk factors.

A qualitative investigation with a standardized interview [4] rendered indicators for psychological disturbances on the basis of harassment through the social environment. As a result this often leads to avoidance

behavior and social retreat, which then leads to interference of the quality of life. Publications uniformly report about a positive influence of the surgical correction of the deformity on the quality of life of these patients. In general they estimated themselves more happily after surgery, felt less tired and the contact with the other gender was facilitated [17]. In the explorative investigation of Roberts et al. [14] the improvement of the quality of life appeared in the subset aspects “satisfaction”, “physical and psychological sense of well-being”, “social integration” and “self-confidence”. Krasopoulos et al. [11] could substantiate that after corrective surgery the quality of life of the patients clearly improved and the self-esteem rose significantly. This was seen in increasing participation in social activities, in rising self-confidence and in the arrangement of social relations. This again led to an increase of self-confidence of the body, the subjectively perceived acceptance of the body through others, the satisfaction with the whole physical appearance and shape (not only the thorax region), the subjective health experience as well as the physical efficiency.
These carried out psychological studies show that a funnel or keel breast deformity can substantially lead to psychological disturbances. Thus Einsiedel and Clausner [6] propagate for *all* patients – irrespectively of the medical severity of the deformity – an *additional supportive psychotherapeutic support*. In their investigations they found out namely that after surgical correction but without therapeutic aid neurotic and inadequate coping strategies increase with rising age.

3.6.2 Psychological test diagnostics

According to Suita et al. [17], in most cases aesthetic-psychological reasons indicate the decision for surgery; however, according to the state of knowledge no comprehensible psychological indication criteria exist so far. Due to the results of available publications it is indeed helpful to measure the following psychosocial dimensions in the patients by psychological questionnaires and interviews.

– Psychological symptoms.
– Quality of life.
– Self-worth and self-confidence.
– Social behavior.
– Body experience.

Numerous sufficiently valid and reliable questionnaires are available in different languages for the acquisition of these dimensions; however, a listing and description of all available instruments would go beyond the scope of this contribution.
Exemplarily and to clarify the extent of a survey, at this point we would like to describe, hence, the psychological diagnostics in the Department for Medical Psychology at the Innsbruck Medical University.
With the patients and as a matter of routine in the phase of the medical preclarification, a detailed bio-psycho-social anamnesis is collected to attain a clear view in respect of the biographical effects of the funnel or keel breast deformity. Potential diagnoses resulting from this interrogation are documented according to the ICD-10 classification system. In addition, a test-psychological investigation is carried out with the following instruments.

3.6.2.1 Brief symptom inventory (BSI)

The “Brief Symptom Inventory” (BSI) [8], which represents a short form of the Symptom-Check List SCL-90-R, collects the subjectively felt psychological burden during the last 7 days. This self-judgment inventory encloses 53 questions (items) which are to be answered on a 5-stage Likert scale from “not at all” (0) to “very strongly” (4). With the BSI nine symptom dimensions and three global identity values can be calculated (somatization, obsessiveness, unsteadyness in social contacts, depression, timidity, aggressiveness/hostility, phobic anxiety, paranoid thinking, and psychotizism). Three global characteristic values (Global Severity index (GSI), Positive Symptom Distress Index (PSDI) and Positive Symptom Totally (PST)) offer a survey over the psychological burden, because they embrace all 53 Items. The GSI measures the basic psychological load, the PSDI measures the intensity of answers in which burden is existent. To fill in the BSI forms it usually takes 8–10 min. The internal consistency (Cronbach’s α) of the single scales lies between $r = 0.39$ and 0.92 and retest reliability reaches from $r = 0.68$ to 0.93. The construct-validity could be proved by very high correlations between the BSI and SCL-90-R scales.

3.6.2.2 Short form 36 health survey (SF-36)

The SF-36 health questionnaires [2] cover the subjective health independently of the state of health from the viewpoint of the affected persons. The subjective health or health-related quality of life is a multidimensional construct, which encloses the physical constitution, social relations, the functional competence and the psychological condition of the interviewees. The questionnaire is suitable for healthy ones as well as for diseased people (somatic and psychological) above the age of 14. There also exists an interview form of the questionnaire, a version for the extrinsic judgment and a short form (SF-12). SF-36 consists of 36 Items which capture 8 dimensions of the subjective health ("physical effectiveness", "physical role function", "physical pains", "general health perception", "vitality", "social effectiveness", "emotional role function", and "psychological well-being"). With the aid of 8 SF-36 subscales it is possible to measure the physical and psychological health of the patient at a higher abstraction level (physical and psychological sum-scale). The answer categories of the items vary from easy binary ("yes–no") up to 6-stage answer scales, average treatment duration amounts 10 min. The internal consistency (Cronbach's α) of the subscales lies between 0.57 and 0.94. The convergent and discriminant validity as well as the sensitivity of the procedure was checked in several investigations successfully.

3.6.2.3 Inventory of interpersonal problems (IIP C)

The "inventory of interpersonal problems" [10] detects the self-assessment of interpersonal problems in interaction with other people. The questionnaire was developed on the basis of interpersonal personality models to enable collection of subjectively felt problems in these interactions. The IIP is applicable for psychologically and physically affected as well as for healthy adults, available as a long form with 127 and an abridged form with 64 Items (IIP C). The realization of the long version claims approximately 15–20 min, of the short version 10–15 min. The answering is carried out on a 5-stage Likert-scale from "not" (0), "a little" (1), "average" (2), "fair" (3) to "very much" (4). Using the 5-stage Likert-scale, 8 scales are measured ("autocratic/dominant", "quarrelsome/competing", "unfriendly/coldly", "introverted/socially withdrawing", "unconfident/submissive", "usable/compliant", "provident/friendly" and "expressive/intrusive"). Measured internal consistency (around 0.50) and retest-reliabilities (0.81–0.90) of the inventory are estimated as sufficient. A sufficient validity in a series of studies, in which the IIP differentiated unambiguously between different clinical diagnosis groups, turned out to be satisfying.

3.6.2.4 Questionnaire for the judgment of own body (FBeK)

The "questionnaires for the judgment of the own body" [16] serves for the ascertainment of the subjective body pattern and body perception. A total of 59 Items are in the first person formulated as a statement and can be answered with "is right" or "is not right". From that four factors can be determined:
"Attraction/self-confidence" (describes the satisfaction with own body and the joy in the own body), "accentuation of the physical appearance" (describes the special stress of the appearance and the joy in the employment with the own body), "insecurity/concern" (describes a rather hypochondric burden of the own body) and "bodily sexual disrelish" (describes sexual discontent or malsensations). The answers to the questionnaire usually last about 10 min, it was primarily developed for adults; however, it can be used in youngsters too. Although this body questionnaire was primarily conceived for psychosomatic research, it has proved itself of value particularly in the clinical-psychotherapeutic area. The internal consistency (Cronbach's α) lies between 0.69 and 0.85 and the Split Half coefficients (to Spearman-Brown and Guttman) between 0.70 and 0.82. Convergent and differential validity of the questionnaire could be proved in numerous studies.
The comprising data from the anamnesis and the test diagnostics result in a comprehensive view with respect to the biography and the determinant psychosocial effects.

3.6.3 Conclusion

In the sense of an interdisciplinary approach and on the basis of the present scientific publications with results regarding the psychosocial effects, a psychological screening should routinely be carried out in all patients with thoracic wall deformities. The synopsis of the data then provides sufficient information about the degree of the psychosocial impairment and the degree of expectations concerning the surgical intervention. The anticipation of the patient about a positive result following surgery with an essential improvement of the quality of life usually is very high. Thereby it must be emphasized that an exclusive operation cannot master these expectations of the patients, as long as serious psychosocial impairments exist or restrictions during a very long period were present. The deficits experienced, possibly lasting for many years, thus correlating to permanently trained experience, naturally left behind traces in the corresponding neural areas in the sense of the theory of the neural plasticity. These cognitive and neural representations must of course be restructured, that is substitution with other experiences. This is the reason why in some patients additional psychotherapeutic aid is also required. As a result of all these conditions the team of therapists has the task to provide the patient with realistic estimations regarding the anticipated physical and psychosocial changes.

The challenge of the future will be to generate concrete cut-off values out of the test-psychological investigation to identify highly stressed patients, who can profit from the operation but with subsequent additional psychotherapy within the psychosocial environment.

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Aspects of indication setting for selection of individual therapy and informed consent

4

Anton H. Schwabegger

The degree of abnormality that is determinable by clinical and laboratory examination is not necessarily proportional to the symptoms the patient has.

R. Ravitch

The type of the surgical correction, which finally will be applied in the correction of a funnel chest deformity occasionally depends there from, to which specialty physician the patient is referred. While thoracic and pediatric surgeons rather will propose the repositioning intervention of the thoracic wall, plastic surgeons might preferably apply different techniques of tissue replacement. Such a pre-selection is depending on the facts, which surgical spectrum and know-how is available at the respective institution. However, just such a course of selection does not represent an optimal selection of treatment that meets the requirements of the patient bearing the aesthetic or functional burden of deformity. Preferably an interdisciplinary discussion of all cases would be of advantage for the concerned patients, but rather not a selection of a procedure, which is just manageable in a singular institution [9]. Being based on the interdisciplinary pool at knowledge and experiences that usually and of course is more comprehensive ("comprehensive care") than a singular expert's opinion, the appropriate choice of treatment or, in selected cases even refusal of any surgical therapy has to be made.

If one considers the WHO definition, after that health means not only the lack of illnesses, but rather also means the condition of a subjective well-being, the surgery of the thoracic wall deformities can improve the mental health of an affected patient. Even if such surgery appears to be performed due to pure aesthetic reasons only, in the sense of intended well-being and with serious indication setting, it may be regarded as a true curative intervention. Likewise in many subdomains of plastic and reconstructive surgery, there hardly exist standards for the correction of the congenital thoracic wall deformities, because continuous developments and variations in the setting of indication take place for the purpose of technical improvements and benefit of experience. Rather than different techniques applied in other deformities, the correction of pectus carinatum is treatable with fundamental standard methods, such as techniques developed by Ravitch in 1960 and Robicsek in 1963 [12, 13]. The indication for the correction of such a keel deformity from a pure (medical) point of view, while lacking of functional somatic deficits, almost always remains an aesthetical indication. However, upon thorough examination the indication for corrective surgery in such cases may almost always be based on accompanying psychical alterations (Chapters 3.6 and 6.3), especially in puberty and adolescence.

In the pectus excavatum deformities on the other hand minimally and minor invasive methods stand in contrast to invasive methods of interventions and furthermore because of a wide range of variety of the extent of a deformity the selection procedures for optimal treatment must be regarded more differentiated. The indication herewith ranges from functional impairment, therewith unambiguously medically indicated [3, 7], until a pure aesthetical indication with predominantly psychological strain but low somatic issue [6]. Apart from a special purely conservative procedure, like the vacuum bell in the funnel deformity (Chapter 6.8) and the chest brace in the carinatum deformity [8] nowadays the palette of therapeutic surgical options is rather diversified and apart from rather simple approaches with silicone implants [9] still rapidly developing [1, 5]. The possibilities of combination of indication setting with many surgical methods are multitudinous and therefore need clearly defined selection criteria for the choice of an optimal procedure for any individual deformity and patient, however, even taking into consideration the necessity of an accompanying psychotherapy pre- and postoperatively (Chapter 3.6).

On the basis of such less medically indicated corrections, the special problematic nature of these predominantly aesthetic corrections therefore comes along with the absolute necessity of a particularly detailed informed consent. In many countries, the jurisdiction demands that, the less an indication is given for a medical treatment, the more exactly and above all more differentiated must be informed about the treatment method of choice or its alternatives. Special and typical risks and potential complications as well as common risks, follow-up accompanying treatment and prognosis of the remodeling intervention must be enlightened, clarified, and well documented. Because many of these interventions in many countries are regarded as a purely aesthetic interventions (e.g., silicone, cartilage chip, lipofilling; Chapters 6.5, 6.6.1 and 6.6.2), particularly these corrections, that are carried out predominately upon special request of the patients but are lacking a distinct medical indication, require a comprehensive and intensive clarification prior to consent and intervention. Especially these patients with only a minor extent of deformity place however very high and often even far super-elevated aesthetic claims at the correction or removal of a minor flaw. The feasibility of an idealized and high-stylized desired result in such patients is often not at all questioned, thus a guarantee for a treatment success then on the basis of the return service by payment is demanded. Especially such patients with unfulfilled expectations then tend to contest the dispute way to the court, even when these expectations correspond to unrealizable und unrealistic ideas. Especially within such situations and subsequent juridical procedures a complete and meticulously documented clarification is of paramount importance for both the physician and the patient with his or her misconception. Under these delicate aspects the intuitional conversation with the patients appears especially important, serving as a prophylaxis to misunderstanding or unbalanced interpretation of expectations, in a minor likewise important as with its parents.

In no case, likewise in aesthetic surgery, promises over a treatment success or guarantee for a desired shaping shall be offered. Above all in asymmetrical deformities of the anterior thoracic wall, the expectations with regard to the realization of a symmetrical result may usually be set very high, but due to anatomical and biomechanical conditions as well as technical limitations are yet hardly to satisfy to date. Where technical feasibility borders at its limits cannot be brought into harmony with the highly elevated expectations of the patients, the consultation of professional psychological aid (Chapter 3.6) through specialists is strongly recommended [2]. Such specialists however should be familiar with the special syndromes of thoracic wall deformities in particular and likewise with the psychological aspects in aesthetic surgery in general.

Long-term alterations such as growth disturbances following too early interventions or late adverse biomechanical effects also are matter of concern for experienced physicians and must be subject to dialogue within informed consent [3, 4, 10, 11, 14].

Chest corrections with elevation of the anterior thoracic wall should be carried out only in clinics with extensive experiences and a widespread repertoire of different treatment methods, a high operation frequency and corresponding routine of the surgeons. Only when these requirements are fulfilled, the intervention is relatively secure and can be performed with a high success rate and an improved functional and/or aesthetic outcome.

To the prerequisites of dealing with thoracic wall deformities also belong to experiences with adequate interdisciplinary diagnostics and follow-up treatment, especially educated and trained physiotherapists and care personnel as well as the availability of an intensive care unit on demand in the situation of emergency complications.

With regard to rare, but possible fatal complication such as heart, lung, or large vessel injuries especially during the MIRPE (Chapter 6.3) or MOVARPE (Chapter 6.4) technique, the intervention should be carried out only in central clinics, in which open heart and lung surgery is commonly performed and immediately available.

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Therapy

5

5.1 Basic surgical techniques with special considerations on skin incisions

Anton H. Schwabegger

While in earlier decades skin horizontal incisions extending up to 30 cm with resulting unsightly scars were not uncommon for surgical access across the anterior thoracic wall and décolleté (Figs. 1 and 2), nowadays incisions and therewith resulting scars of 3–10 cm usually suffice for the different techniques of correction applied.

Fig. 1. Large horizontal scars crossing the midline, funnel chest surgery performed four decades ago

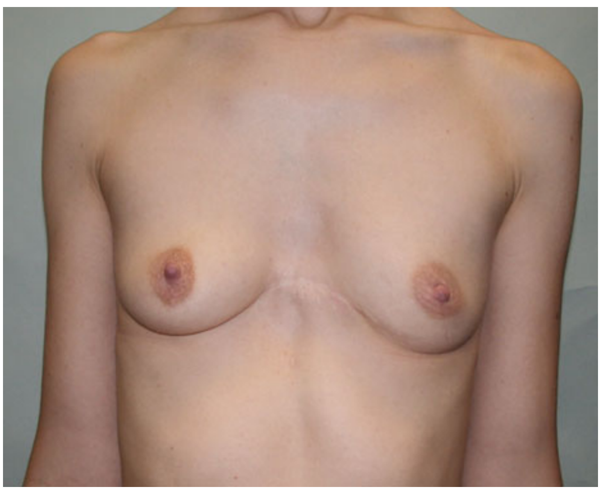

Fig. 2. Well-hidden scar at the inframammary crease, also resulting from funnel chest surgery decades ago, but with still very well visible stain within the décolleté

The conventional surgical accesses in male patients with keel or in funnel chest deformity usually are set centrally above the sternum. Although no evidenced data are available so far, which kind of incision, whether vertically, horizontally or with other distinct design like lazy-S or W-shaped (Figs. 3–6) will lead to development of the slightest and unrecognizable scar (Chapter 12.2). Particularly the presternal region is well known and accused to development hypertrophic scars or even cumbersome keloids [4], even if one follows the criteria of skin incision along the relaxed skin tension lines (RSTL). However, within our own series of patients operated on keel chest deformity, we have observed

Fig. 3. Horizontal inconspicuous scars placed along the RSTL in a 27-year-old man who received a horizontally placed custom-made silicon implant into lower arch rib depressions, via the left incision even a minor gibbus of the fourth and fifth left rib cartilages parasternally could be resected simultaneously

Fig. 4. W-shaped central access for sternum elevation during the MIRPE-procedure, scar fortunately remained soft 4 years after the initial surgery. However, nowadays a scar within the submammary crease would be preferred

Fig. 5. Horizontal scar at the right side, this small incision, enabled multiple cartilage resections of the second to the sixth rib in an asymmetrical keel chest deformity without sternum malrotation

only a few partial hypertrophic scars but no keloids. Above all, in the correction of keel chest deformities, the continuous postoperative compression of the scar by the keel chest brace (Chapter 7.4) over a period of 6–8 weeks may distinctly be responsible to work against such disadvantageous development of hypertrophic scars (Fig. 7) or keloids [6, 8]. On the other hand, relative skin excess is generated through the skeletal reduction of the cavity in the funnel or the convexity in the keel

Fig. 6. Arched incision for access to microvascular sternum turn-over flap (Chapter 6.6.5) in extensive pectus excavatum

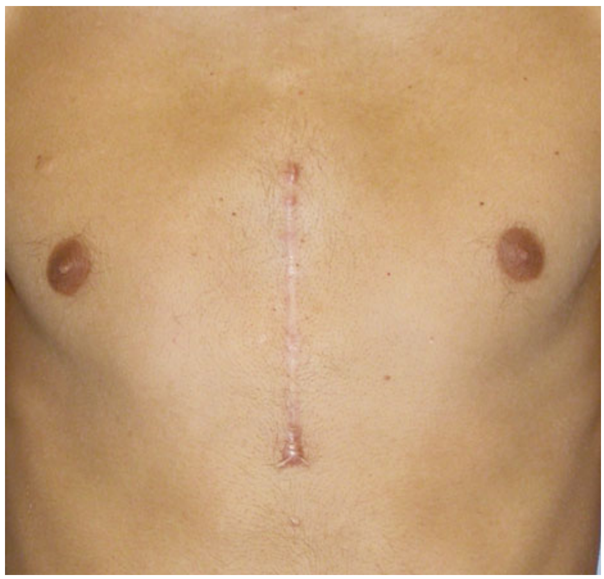

Fig. 7. Vertically placed scar after extensive keel chest correction including horizontal sternum osteotomy. Notice the flat and smooth part of the scar, exactly where the silicon pad of the keel chest brace (Chapter 7.4) was placed for 8 weeks. The scar extending the area of the pad cranially and caudally, thus left without compression developed hypertrophic zones

chest deformity, which relaxes the wound seaming from any tension. Therefore the causing agents namely wound tension, which is responsible for the excessive collagen production are omitted and therewith any hypertrophic scar development is circumvented too, hypothetically. However, it is very well known clinically but still without high-level evidence data, that compression therapy on hypertrophic scars and keloids

Fig. 8. This patient had extensively deep pectus excavatum operated at the age of 15, using a modified approach with bilateral submammary incisions remodeling of sternum and adjacent ribs, additionally implantation of a pectus bar. Situation 1 year after pectus-bar explantation at the age of 17 and moderately grown breasts shows slight displacement of the scars to their undersurface, however well concealable by bra

Fig. 9. Definition of the submammary crease laterally during MIRPE surgery to place the incisions exactly there. Preferably these incisions should already be marked in standing position prior to surgery, to include the gravitational effect on the tissue, thus presenting with a different location of the lateral and submammary crease

Fig. 10. Lateral aspect of a 17-year-old female with inconspicuous scars for the MIRPE-procedure exactly placed into the lateral breast fold

definitely benefits their outcome and aesthetic appearance [2, 6].

In female patients the skin incisions are placed into the submammary crease, similar to the surgical access in reduction mammoplasty (Fig. 8), therefore they are very well hidden in a frontal aspect. The lateral incisions in the MIRPE-technique are also placed into or along the extensions of the submammary crease, where they ideally can be concealed by bra (Figs. 9 and 10).

In the correction of the pectus carinatum or pectus excavatum deformity, scars in the median line must be avoided, i.e., the female décolleté should remain untouched by incisions in any case. This may impede the access to the presternal region, especially when a horizontal osteotomy of the sternum is necessary. However with the experiences in reduction or augmentation mammoplasty, this problem can be handled thoroughly well. A specially angled blade and oscillating saw applicable especially for that submammary access to reach the sternum (Fig. 11) is described in Chapter 9.

For the case that simultaneously to a chest remodeling access in female adults also mammary implants were necessary, the correct planning of scar positioning may become cumbersome. Herewith it has to be considered

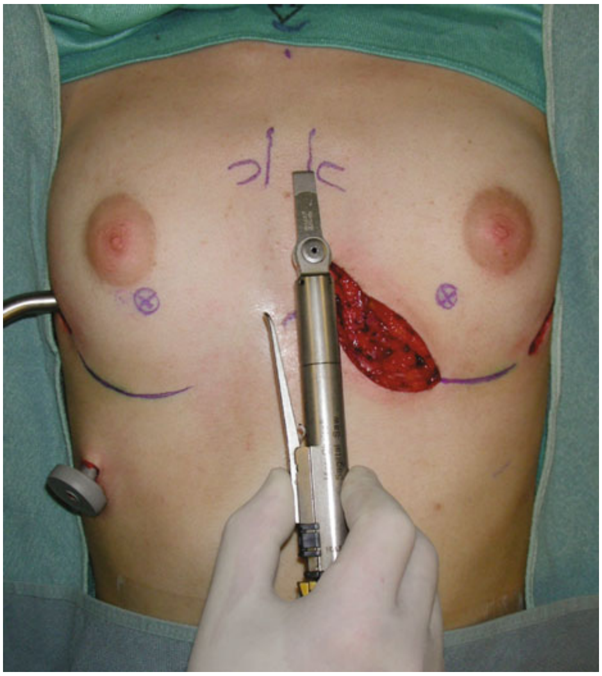

Fig. 11. Submammary access for horizontal sternum osteotomy in patients with either keel or in this particular case funnel chest deformity, enabling subcutaneous approach of the oscillating saw with an angle blade to the desired level of the osteotomy

that through the implant expansion of the mammary gland tissue also skin distension will occur, leading to a dislocation of the scar from the inframammary crease further cranially and therewith scars come to lie at the underside of the breast skin (Fig. 8). Therefore comprehensive experiences with the augmentation mammoplasty procedures are required in this simultaneous correction to optimize the aesthetic outcome, which is of major importance in the female patient particularly. Disfigured and prominent, usually only aesthetically disturbing rib arches, as far as they not can be corrected by any central access, or are subject of solely isolated treatment, are resected through separate incisions, exactly following the lines of Langer or RSTL [1, 3, 5, 7]. The wounds of all incisions are closed by means of intracutaneous running sutures only. Because usually a relative surplus (Fig. 12) of skin emerges by reduction of a cavity or a convexity, absorbable subcutaneous sutures for the relief of skin tension are unnecessary, because they are subject to produce localized necrosis of subcutaneous fat with ensuing development of scar contraction in the subcutaneous fatty tissue. Such adherent and sunken scars (Fig. 13) then generate so-called unaesthetical “eye-catchers” and persistent-

Fig. 12. Surplus of tissue without excision of excessive skin after correction of pectus arcuatum causes skin folding

Fig. 13. Retracted scar in this slightly obese patient, potentially caused by inappropriate suture technique with subcutaneous fat necrosis and ensuing tissue loss with scar formation subcutaneously

ly direct the attention often more on itself than on the previously existed deformity.

The aesthetically sensitive zone of the female décolleté should, if possible,remain surgically untouched in any case and therewith taboo for any scarring. Nearly all surgical accesses to the sternum, to the parasternal rib region even highly up until to the second or third rib are manageable out from an access along the submammary crease. On the one hand, connecting scars horizontally between both breasts tend to develop scar contraction and breast distortion. On the other hand, and in contrast to scars along the submammary crease, such horizontal scars crossing the midline are well visible (Figs. 1 and 2) lifelong for others and for the patient when looking downward or into the mirror. Thus the stigma of deformity would only have been replaced through the stigma of a well visible scar then, which would mean no definite improvement of body image and self worth.

With the implantation of custom-made silicone implants, the scars at the chest should preferably come to lie distant from the median line, again to circumvent the zone of probable scar hypertrophy or keloid formation, and furthermore to avoid (Figs. 10 and 11 in Chapter 6.5) stigmatization of the central body aspect for aesthetic reasons above all. In female patients the endoscopic implantation of custom-made silicon blocks, which are brought in through a periumbilical incision [4], represents an interesting alternative to avoid scars in the breast area. For sure, the endoscopic preparation of the recipient pocket via the distant access is a more skillful and time-consuming procedure than a direct access, but the aesthetic benefit especially for young and female patients may be impressive.

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5.2 Positioning of patients during surgery

Anton H. Schwabegger

In almost all operations for the correction of thoracic wall deformities, the patient is positioned supine on a flat operation table, and he/she will retain this position during the entire time of the intervention. A particular exception is the correction of an asymmetric female breast as far as this concomitant deformity is corrected simultaneously in the same operation with the thoracoplasty. Herewith the torso of the patient usually will be erected up to 45° to estimate the natural form and ptosis of the untouched contralateral breast, so that the breast to be operated can be adapted optimally to that relevant shape. An other exception from pure supine positioning is necessary in harvesting of a latissimus dorsi flap for the correction of a Poland-syndrome exemplarily, pedicled or transplanted as a micro (neuro)vascular flap (Chapters 10.1 and 10.3). In such cases the position of the torso must either be solely elevated [2] or entirely turned to a lateral decubitus position to enable dissection of the latissimus dorsi flap as a partial or entire flap to be transposed or transplanted. A further exception is the surgery for pectus bar explantation in children because herewith and in contrast to the more laterally extended shape and overall rigidity of the thorax in adults, the pectus-bar needs not to be bent back to a straight shape, but rather can be extracted from the thorax along the almost uniformly round-curved shape of the implanted pectus-bar, with the child positioned laterally [1] (Chapter 6.9).

5.2.1 Positioning during funnel chest surgery

In the correction of the funnel chest deformity, utilizing either the MIRPE, similar endoscopically assisted semi-open or other techniques, the arms are adducted (Fig. 1) at the shoulder and bent in the elbow joints to cranial around 90° to gain sufficient space for the manoeuvres of the endoscopy equipment (Chapter 5.4 with Figs. 1 and 2 and Chapter 6.3 with Figs. 5 and 8). In addition also the placement of a pillow underneath the thoracal spinal column is recommended to elevate the thorax to some extent and therefore to expand the overall maneuverability of the thoracoscope (Figs. 1 and 2). Particular care must be taken to a correct positioning of the arms, as

Fig. 1. Patient positioned for the MIRPE technique, both arms are abducted and bent in the elbow joint in order to gain sufficient space for maneuvers for the endoscope, the tunnelizer and the pectus-bar implantation tools

Fig. 2. Intraoperative situs with arms adducted and bent at the elbow joint in an adult tall male, undergoing pectus excavatum deformity correction by a combined approach

brachial plexus lesions, temporarily or even permanently may result from over-elevation or unintended retroversion potentially lasting for hours in the cases requiring complex remodelling.
For the particular cases that surgery is performed according to the conventional technique of Ravitch exemplarily and no additional lateral approach to the thorax is mandatory, both arms will be adducted to the torso for the purpose of relaxation of the pectoralis muscles. Even the origins of the rectus abdominis muscles are advantageously relaxed by that setting, due to slackening of the entire thoracic cage, consecutively to relaxation of the Pectoralis major muscles.

Fig. 3. Patient positioned for keel chest correction, surgery is already completed. Both arms remained adducted in order to relax the pectoralis major and rectus abdominis muscles for either transmuscular or submuscular access

5.2.2 Positioning during keel chest surgery

In contrast to the positioning in the (minimally) invasive funnel chest surgery in the correction of the keel chest deformity both arms should be adducted to the torso in order to relax the Pectoralis major muscles (Fig. 3). By that means the muscle preparation is facilitated, independently if either the muscles are conventionally elevated as flaps from the skeletal thorax, or a muscle split technique is applied to approach the rib cartilages [3]. Moreover, through this positioning, i.e., the adduction of both arms, the lateral tension of muscles of the shoulder girdle on the skeletal chest is minimized, thus and as a consequence the rectus abdominis muscles inserting on the lower rib arches are also relaxed. These abdominal muscle insertions notably also have to be dissected or split longitudinally for the access to the rib cartilages along the inferior costal arch.

5.2.3 Positioning during surgery of other deformities

The positioning in other thoracic wall deformities depends on the necessity whether thoracoscopy is necessary or not. In all interventions, that come along without endoscopic access, the positioning with adducted arms is recommended in order to relax the thoracic cage and thus to facilitate the surgical preparation of the anatomic structures especially the muscles, but moreover to enable a proper anatomic repositioning of eventually detached muscle flaps from the sternum and adjacent ribs.

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5.3 Special anesthetic considerations in thoracic wall surgery: epidural anesthesia, lung separation, and postoperative analgesia

Günter Luckner, Gottfried Mitterschiffthaler, Martin W. Dünser

5.3.1 Introduction

According to North American data, the minimally invasive reconstruction of pectus excavatum (MIRPE) procedure is performed at an average patient age of 12.4 years [1]. In Europe, the corresponding mean patient age varies between 17.6 ± 5.8 and 15.1 years [2, 3]. Therefore, most anesthesias for the MIRPE procedure need to be performed in pupils and adolescents. In majority of the cases, the MIRPE procedure, as originally described by Nuss or as a combined semi-open approach in adults, is performed for idiopathic thoracic wall deformity, while some patients present with iatrogenic thoracic wall deformity (e.g., following cardiac surgery) or deformities secondary to connective tissue disease (e.g., Ehlers–Danlos or Marfan syndrome). Most patients with thoracic wall deformities do not exhibit substantial lung function deficits [1]. A history of exhaustion after limited exercise should prompt preoperative spirometric testing. Most commonly, those patients who show decreased lung function suffer from a restrictive ventilatory pattern.

Preoperative cardiologic workup is indicated only in symptomatic patients [4]. If thoracic wall deformity is present secondary to cardiac surgery, recent cardiologic reports should be checked and the administration of antibiotic prophylaxis considered to prevent endocarditis.

Patients with a keel chest deformity also undergo thoracoplasties with similar invasiveness to the skeletal structures thus suffering from postoperative pain. While in these patients, in contrast to the MIRPE or similar procedures, a conventional tracheal intubation is sufficient, postoperative pain control is necessary likewise to funnel chest surgery.

5.3.2 Pre-anesthetic consultation

During the pre-anesthetic consultation, a detailed medical history is taken. It is especially important to explore a history of abnormal bleeding or allergies. In underage patients, written informed consent is obtained from both patient and legal guardians. Patient information and medical history must be meticulously documented, especially concerning risks and benefits of invasive procedures such as epidural catheter and central venous line placement. In light of its paramount importance in postoperative pain management for major surgery at funnel and the keel chest deformities, placement of an epidural catheter should always be scheduled as long as there are no contraindications [5]. Awake puncture is considered feasible from age (10–)12 years, but should not be forced upon the patient, and should always be discussed with both the patient and his legal guardians. Adequate postoperative analgesia can also be achieved with continuous intravenous infusion of opioids, provided that appropriate monitoring is available [6].

Also discussed are airway management using double lumen tubes (DLTs), and the technique of ventilatory lung separation [7, 20, 21, 26]. Additional compulsory issues discussed during the preoperative consultation are specific invasive procedures such as a central venous catheter and arterial line, which are mandatory at our institution considering the potential complications for pectus excavatum surgery [8, 9]; these issues may be omitted for pectus carinatum surgery.

Erythrocyte concentrates should be readily available for transfusion. At our institution, we routinely have two units on hand. Routine preoperative laboratory tests include blood count, standard coagulation markers (partial thromboplastin time, prothrombin time), and serum biochemistry, but there is no evidence to support the use of these laboratory tests.

Postoperative care should be scheduled for the first 24 h to assure optimal monitoring, pain therapy, and mobilization.

Patients are free to ingest clear fluids until 2 h preoperatively, and two sites for venous puncture are primed using a eutectic mixture of local anesthetic (EMLA®) plasters.

5.3.3 Anesthesia

On arrival in the operating room, the patient is welcomed by the anesthetist and positioned on the operating table. The sensitivity of young patients undergoing this procedure is to be dealt with tactfully.

5.3.3.1 Thoracic epidural anesthesia

Thoracic epidural anesthesia is a segmental blockade of autonomous and somatic nerves with an upper and a lower sensory level. These levels need to be adapted for the particular surgical procedure. The advantages of thoracic epidural anesthesia do not substantially differ for adolescents and adults [10] and include excellent intraoperative analgesia without needing high-dose opioid administration, and the postoperative provision of continuous epidural analgesia [11–13].

General considerations for the loss-of-resistance method used at the mid-thoracic level

Good knowledge of vertebral column anatomy is essential in the performance of epidural anesthesia. The distance between the ligamentum flavum and the dura is approximately 2–3 mm in the higher thoracic region, and increases to 3–5 mm in the lower thoracic region. Spinous processes are angled almost horizontally at the cervical levels, and downward between 40° and 70° in the mid-thoracic region (Fig. 1). This angle, which may interfere with the median approach to epidural catheter placement, becomes less pronounced toward the lower thoracic levels [14].

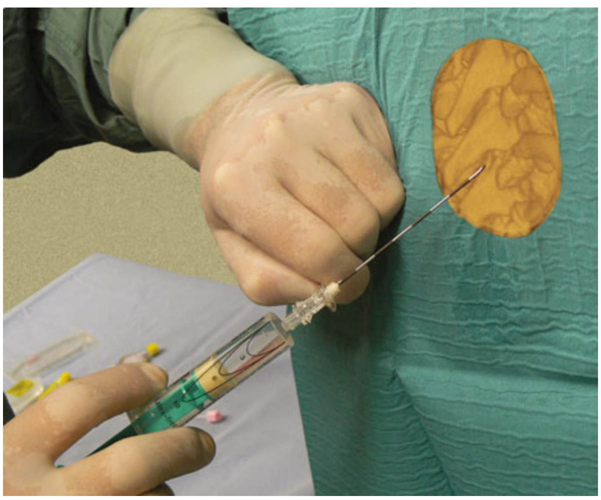

Fig. 1. The left hand is used to advance the needle and control its depth of penetration below the skin. The right hand is used to maintain pressure on the loss-of-resistance needle. Spinous processes are angled downward between 40° and 70° in the mid-thoracic region

The puncture level is determined primarily by evaluating osseous landmarks. Most importantly, a line connecting the lower scapular angles indicates the level of the seventh thoracic vertebra.

Before puncture, the skin and subcutaneous tissues are infiltrated with local anesthetic to permit pain-free epidural puncture. To ensure minimum discomfort for the patient during epidural puncture, it is important to allow sufficient time for the local anesthetic to take effect.

In children in whom an awake puncture is planned, the puncture area can be prepared using a local anesthetic cream or plaster (EMLA®).

Positioning and approach

Epidural catheter placement is performed in the monitored patient (pulse oxymetry, electrocardiogram, and non-invasive blood pressure) only after venous access has been established. Puncture is performed in a sitting position, with the patient bent forward, his elbows resting on his thighs, and his head bent down. This position helps bring the thoracic spine into a more lordotic position, facilitating epidural access.

The patient should be made aware that sudden movements may be detrimental. To ensure the patient's immobility, an anesthesia nurse supports the patient from anterior.

The puncture set for thoracic epidural anesthesia and the sterile gown for the anesthetist performing epidural anesthesia are prepared before the patient is brought into a sitting position.

In young schoolchildren under the age of 10–12 years, we perform the epidural access under general anesthesia in the left lateral decubitus position while gently flexing the patient's vertebral column.

The paramedian approach can be performed at all thoracic levels, whereas the median approach can be technically difficult or impossible between the levels of the third and ninth thoracic vertebra due to spinous process configuration. For this reason, we primarily use the paramedian approach.

Paramedian approach

We mark the central points of two adjacent interspaces between spinous processes, and draw a line connecting these points. From the mid-point of this line, the puncture site is found by moving 1(–2) cm laterally. The epidural puncture is performed at 15° toward the median sagittal plane, with the needle directed 50° cranial. The angles cited here are only approximate values that need to be tailored to the particular patient by the experienced clinician.

About 1 cm below skin level, the needle's stylet is removed, and the loss-of-resistance syringe filled with normal saline is connected to the needle. The left hand is used to advance the needle and control its depth of penetration below the skin. While the thumb and second finger of the left hand are used to advance the needle, the third to fifth fingers stabilize the needle against the patient's back. The right hand is used to maintain pressure on the loss-of-resistance needle, but does not assist in needle advancement (Fig. 1). The distance from skin to epidural space shows considerable interindividual variability, and is about 3–6 cm in adults. Therefore, loss of resistance may be encountered only after a few centimeters.

In children, the distance from skin to epidural space can be estimated using formulas originally devised for the lumbar epidural space. At birth, the distance between skin and lumbar epidural space is about 10 mm, and increases in a linear fashion with age. In older children, Busoni's formula is frequently used [(age in years × 2) + 10 mm)] [15]. It should be remembered, however, that these estimates are valid only for the lumbar epidural space, and are only rough indicators for the distances encountered in the thoracic region.

When the ligamentum flavum is penetrated, a sudden loss of resistance is perceived, both to needle advancement and to fluid injection.

On entering the epidural space, the catheter is threaded approximately 4–5 cm into it.

Before meticulous fixation of the catheter, intravascular or spinal misplacement should be ruled out. This is done by excluding passive reflux of blood or cerebrospinal fluid via the catheter and by attempting active aspiration using a small (2 cc) syringe. It should be noted, though, that even if reflux or aspiration of blood or cerebrospinal fluid is absent, intravascular or intrathecal catheter localization cannot be fully excluded.

5.3.3.2 Drugs and dosing for adolescents and adults

The volume of local anesthetic administered determines the number of segments blocked and therefore the longitudinal spread of epidural anesthesia. As an initial dose in adults and adolescents, we apply 1.0–1.5 ml of local anesthetic per segment. In children, we use 0.75 ml/kg body weight.

The concentration of local anesthetic is relevant to block density. Local anesthetic administered at low concentrations (ropivacaine 0.1%, bupivacaine 0.125%) primarily causes sympathetic and sensory blockade, while administration of local anesthetic at high concentrations (ropivacaine 0.2–0.5%, bupivacaine 0.5%) produces sufficient intraoperative analgesia and motor blockade. Local anesthetics used at higher concentrations are therefore beneficial, if intraoperative muscular relaxation is desired in addition to sensory blockade.

In the postoperative phase, small to medium concentrations of local anesthetic are used for analgesia, which should optimally spare motor function.

Addition of opioid to the local anesthetic has a synergistic analgesic effect and therefore permits the local anesthetic dose to be reduced.

The particular local anesthetic and its dosage are decided by the anesthetist performing the epidural puncture. A dose overview of commonly used local anesthetics is found in Table 1. In children, doses should be adapted and maximum allowable doses kept in mind.

As an initial bolus, we use 8–15 ml ropivacaine 0.2% (or bupivacaine 0.25%) plus 100 µg fentanyl. For intraoperative maintenance, we use a perfusion pump working at 6–10 ml/h. The perfusor may be prepared as

Table 1: Local anesthetics commonly used in epidural techniques under combined general and thoracic anesthesia – adults and adolescents (>50 kg)

Anesthetic	Concentration	Initial bolus	Intraoperative	Baseline rate for patient-controlled thoracic epidural analgesia (PCTEA)
Ropivacaine	0.2–0.5%	5–15 ml plus opiate: e.g. sufentanil 1 µg/ml or fentanyl 100 µg	Repeated dose: Half of the initial dose (approximately every 60–90 min) Continuous dose: 6–10 ml/h ropivacaine 0.2–0.5 %	Ropivacaine 0.2% + e.g. plus sufentanil 0.5 µg/ml: 5–7 ml/h; bolus 2 ml; lock-out time 20 min
Bupivacaine	0.25–0.5%	5–15 ml plus opiate: e.g. sufentanil 1 µg/ml or fentanyl 100 µg	Repeated dose: Half of the initial dose (approximately every 60–90 min) Continuous dose: 6–10 ml/h bupivacaine 0.125–0.5%	Bupivacaine 0.175% + e.g. plus sufentanil 0.5 µg/ml: bolus 2 ml; lock-out time 20 min

follows: 50 ml of ropivacaine 0.2% plus 100 μg fentanyl: 10 ml of ropivacaine 1% plus 2 ml of fentanyl (100 μg), plus 38 ml normal saline. This regimen permits 10–12 spinal segments to be blocked.

Spread of epidural anesthesia should be tested preoperatively. Spread beyond C8 causes sensory loss in the small finger, and diaphragmatic function is lost when local anesthetic blocks the fourth cervical nerves.

Safe practice of thoracic epidural anesthesia is assured only when the practitioner has a thorough theoretical knowledge of indications, contraindications, and potential complications entailed [16, 17]. Apparent or potential complications such as signs of epidural space compromise (motor weakness, backache, paresthesia, paralysis, cervical rigor, and headache) should be readily recognized and diagnosed by computed tomography (CT) or magnetic resonance imaging (MRI), and promptly managed. If epidural hematomas are not surgically decompressed within about 6 h, neurologic recovery is not likely. Knowledge of the mandatory intervals between administration of anticoagulants and insertion or removal of epidural catheters is imperative [18, 19].

Anesthetist experience with slow injection of local anesthetic under repeated aspiration after previous administration of a test dose containing adrenalin is also a prerequisite.

5.3.3.3 General anesthesia

After thoracic epidural anesthesia has been initiated, the patient is brought into a supine position. For induction of general anesthesia, we use propofol 2.5–3.5 ml/kg body weight, fentanyl 3 μg/kg body weight, and rocuronium 0.6 mg/kg body weight. Complete muscle paralysis is verified using a peripheral nerve stimulator. Intubating conditions should be optimal when airway management is performed using a DLT, because the risk of injury is considerably increased when this device is used [20] but most of the complications encountered with double lumen tube placement (malposition, tracheobronchial tree disruption, and traumatic laryngitis) can be avoided by multiple repeatedly checkings on tube position and by selection of appropriately sized tubes.

Intubation using a DLT allows for ventilatory lung separation [21]. During the Nuss procedure, the main indication for separate lung ventilation is the optimization of surgical conditions. Experience in device handling and tracheobronchial anatomy are essential to safely perform DLT insertion. We use the PVC Robertshaw model with no carinal hook (Broncho-Cath™ MALLINCKRODT). For thoracoscopic and extrapulmonary surgery, left-sided DLTs are generally recommended. These are available from size 26–41 Ch. The tracheal diameter is considered a good indicator for DLT size [22, 23]. In children under the age of 8–10 years and in small adults, lung separation can alternatively be performed with a bronchial blocker or deliberate left-sided intubation using a conventional endotracheal tube. The smallest available DLT has a size of 26 Ch, corresponding to an endotracheal tube with an internal diameter of 6 mm.

Approximate size of DLT for adults:

- 35 Ch: <165 cm height,
- 37 Ch: 165–175 cm height,
- 39 Ch: 175–185 cm height,
- 41 Ch: >185 cm height, and
- smaller available sizes: 26/28 and 32 Ch.

Patient positioning and DLT intubation: For intubation, the patient's head is brought into a Jackson position, while the entire patient is brought into a 15° reverse Trendelenburg position. The patient's teeth are protected with a gumshield. After laryngoscopic visualization of the vocal cords, the left-sided DLT is advanced into the trachea with the distal tip facing anterior. Subsequently, the DLT stylet is removed, and the tube rotated 90° counterclockwise. Now, the tube is gently advanced until an elastic resistance is encountered. When using a left-sided DLT, this is usually elicited by fixation of the distal bronchial tube in the left main bronchus. Now, the proximal (tracheal, transparent) cuff is blocked first. To estimate depth of tube insertion, the distance between the middle of the clavicle and the carina can be measured on a chest radiograph [24]. The gold standard, however, is fiberoptic verification of DLT position [25, 26]. Introduction of the flexible fiberscope is possible via a universal adaptor (Mainzer Universaladapter®, Rüsch, Waiblingen). To this end, the DLT's tube adaptor is clamped proximally on its tracheal limb, and the bronchoscope is inserted into the tracheal tube. When the tracheal bifurcation and both main bronchi are visualized, the anesthesia nurse blocks the distal (bronchial, blue) tube in the left main bronchus. The tip of the flexible fiberendoscope is positioned in the distal orifice of the tracheal lumen to allow visualization of the DLT (Figs. 2 and 3). The figure shows the correct position of the bronchial (blue) cuff just beneath the tracheal carina in the left main stem bronchus, with unobstructed view of the carina itself. To inspect DLT position, a fiberendoscope with an outer diameter of 3.6 mm is used. This endoscope allows verification of position in DLT down to a size of 35 Ch. For smaller DLT

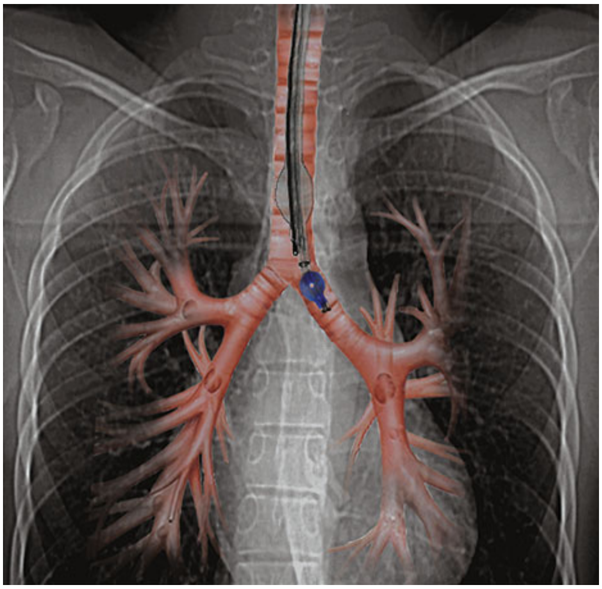

Fig. 2. Depicts fiberoptic verification of left-sided DLT position via the tracheal lumen, with correct position of the tracheal (transparent) and bronchial (blue) cuffs

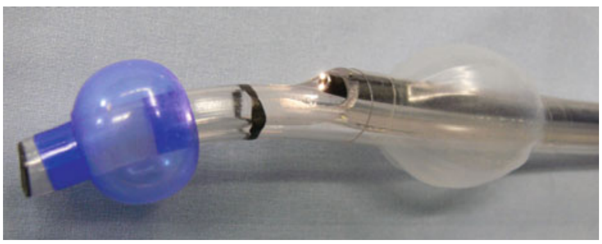

Fig. 3. The tip of the flexible fiberendoscope is positioned in the distal orifice of the tracheal lumen to the DLT

size 26–32, an ultra-thin fiberendoscope with an outer diameter of 2.2 mm should be used. When the DLT is correctly positioned, the inflated bronchial cuff can be seen as a narrow blue crescent on the medial side of the tracheal bifurcation. The DLT should be secured in this position. However, in our opinion, the best method for placing DLTs on first try was first described in 1996 by Ovassapian. For this technique the tube is first advanced only into the tracheal lumen. Then the fiber endoscope is moved through the endobronchial lumen, until the carina and mainstem bronchi are identified. Next, the endoscope is advanced into the proper bronchus and, using it as a stylet, the DLT is slid over the fiber endoscope until the bronchial lumen comes into sight ahead of the tip of the fiber endoscope. Finally, the fiber endoscope is moved through the DLT's tracheal lumen

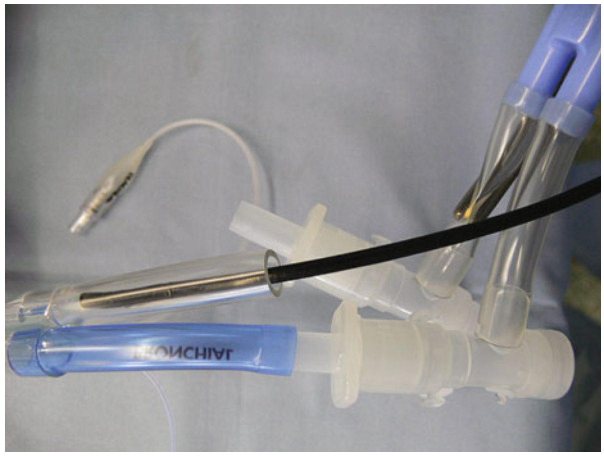

Fig. 4. Introduction of the flexible fiberscope in the proximal limb of the right limb of the tube connector, and disconnecting the distal limb of the DLT

to confirm that the position of the DLT is correct [27]. DLT stability can be enhanced by inserting an additional Guedel tube. To exclude dislocation of the DLT (e.g., herniation of the distal cuff into the trachea), the position should be repeatedly verified, especially after each change in patient position and whenever oxygenation seems impaired.

Immediately before the surgeon enters the thoracic cavity, lung ventilation should be separated by clamping the proximal limb of the left or right limb of the tube connector, and disconnecting the respective distal limb (Fig. 4).

After DLT placement, a central venous catheter may be inserted under ultrasound control via the subclavian or internal jugular vein. A Foley catheter should be placed to monitor urine output during surgery. After cannulation of the radial artery, the anesthetic procedures are completed.

Specific anesthetic responsibilities during surgery include routine verification of patient position, DLT position, and temperature management. Anesthesia can be maintained by continuous infusion of propofol (1%, 30–50 ml/h) and low-dose remifentanil (0.05–01 µg/kg/min), the latter to provide analgesia for positioning and tube tolerance.

Intraoperative fluid substitution is performed using crystalloid solutions. For moderate surgical trauma, approximately 3–5 ml/kg body weight/h is infused. The rate of infusion and the type of solution (crystalloid versus colloid) need to be adapted to surgical progress and the extent of surgical trauma [28] and necessitate continuous communication between surgeon and anesthetist.

During one-lung ventilation, inspiratory oxygen concentration is initially increased to 100%, and later adapted to periodic blood gas analyses. The standard ventilator setting for one-lung ventilation is approximately the same tidal volume as for two-lung ventilation (10 ml/kg). Respiratory rate is adjusted to maintain normal $paCO_2$ [29]. Nitrous oxide is not used during one-lung ventilation or thoracoscopy. At the end of surgery, the patient should be promptly extubated.

5.3.4 Postoperative patient management

Owing to the nature of surgery, pain management is the most important postoperative concern. Continuous epidural infusion of local anesthetic is the method of choice, and the patient should be regularly evaluated for block level, sufficient pain control, and adverse events. Quality of pain therapy is evaluated using a 0–10 Numeric Pain Intensity Scale.

The standard epidural infusion at our institution is 0.2% ropivacaine combined with 100 µg fentanyl (in 50 ml normal saline), administered at 6–10 ml/h.

After the patient is transferred from the postoperative intermediate care unit to the normal ward, epidural infusion is continued in the form of patient-controlled epidural anesthesia (PCEA) under the supervision of the institutional pain service. The epidural catheter should be checked regularly for signs of infection and removed between the third and fifth postoperative day.

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5.4 Video-assisted thoracoscopy (VATS)

Paolo Lucciarini, Anton H. Schwabegger, Thomas Schmid

5.4.1 Introduction

Since the introduction of video-assisted thoracoscopy (VATS), this endoscopic procedure has evolved to a safe, standardized treatment supporting modality also for the minimally invasive repair (MIRPE) of congenital thoracic wall anomalies, namely for the surgical treatment of pectus excavatum deformity, nowadays performed in numerous specialized surgical referral centers.

The minimally invasive placement of one or two individually formed metal struts for reshaping a sunken sternum with its adjacent ribs was initially described by Donald Nuss in 1998 (Chapter 6.3), remarkably after a follow-up period and cumulative experience of 10 years [1, 2]. Concomitantly with the general technical development of video-assisted thoracoscopic equipments and increasing knowledge in thoracoscopic operations, such as minimally invasive apical lung wedge-resections or evacuation of thoracic empyema exemplarily, the MIRPE was developed. Pediatric and reconstructive surgeons likewise [3–5] adopted endoscopic assistance and control of instruments or pectus-bar hardware implantation in pectus excavatum surgery. Its routine use led to a marked reduction of intraoperative complications, which are assumed but not reported along with the first series of the MIRPE interventions. At that time, the pioneer surgeons performed pectus-bar implantation yet without assistance of an intraoperative thoracoscope. Such complications were injuries of lung parenchyma due to pre-existing pleural adhesions with subsequent air leaks and/or hemorrhage, bleeding from intercostal or internal thoracic vessels [6] during forced placement of the metal bars, with shearing and disruption effects on intrathoracal tissue or improper placement of the metal strut with late sequela. In exceptional cases, life-threatening events and fatal outcome had also been described only recently, mostly because of injury to the heart and/or great vessels after uncontrolled hardware insertion or removal maneuver [7–14]. In particular, the assistance of VATS in the MIRPE of adolescents or adults is of paramount importance, since all thoracic wall structures at that age feature much more rigidity than those in children. Because of such restricted mobility of the anterior thoracic wall, narrowed and rigid space for surgical maneuvers due to the immobility of the sunken sternum and adjacent ribs is usually existent in adults, notably in athletic males. Therefore the endoscope is an utterly necessary tool to provide with survey and a safeguard against potential intrathoracal tissue damage during implantation of instruments and final placement of the pectus bar.

5.4.2 Surgical technique

The patient is placed in a slightly overextended supine position with a foamy pad underneath the shoulders for proper exposition of the sunken sternum and deformed anterior thoracic wall. The arms are abducted 90° from the trunk (Figs. 1 and 2) on both sides to improve maneuverability of the videoendoscopic system, with attached camera, hooked light cable and electronic wires. Double-lumen tracheal intubation (Chapter 5.3)

Fig. 1. Positioning of the patient for videoendoscopic access. Arms are abducted with flexion at the elbows to gain sufficient place for both the pectus-bar implantation and the endoscopic maneuvers, postoperative view, notice the double-lumen tracheal tube

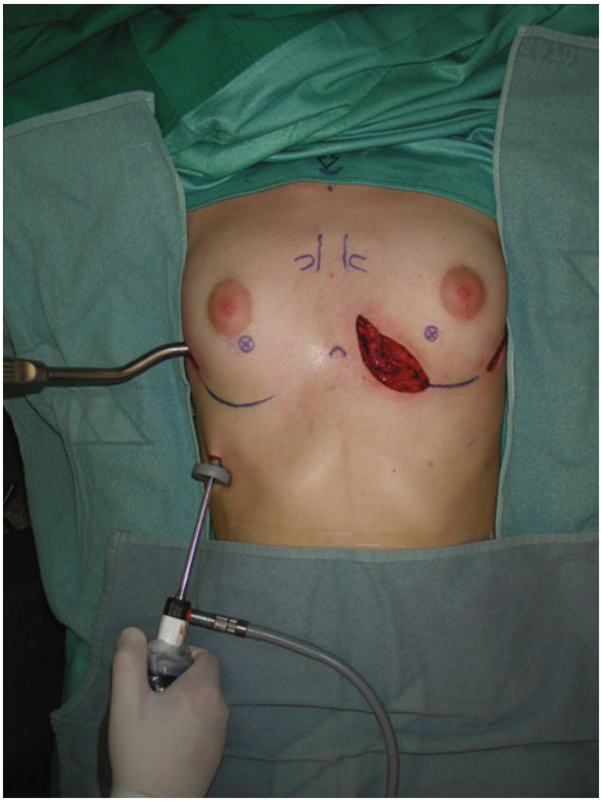

Fig. 2. Intraoperative situation with the endoscope and the pectus-bar flipper in place. This is a modified semi-open approach with thoracotomy and necessary osteochondrotomy in a young female adult with a rigid and deep deformity

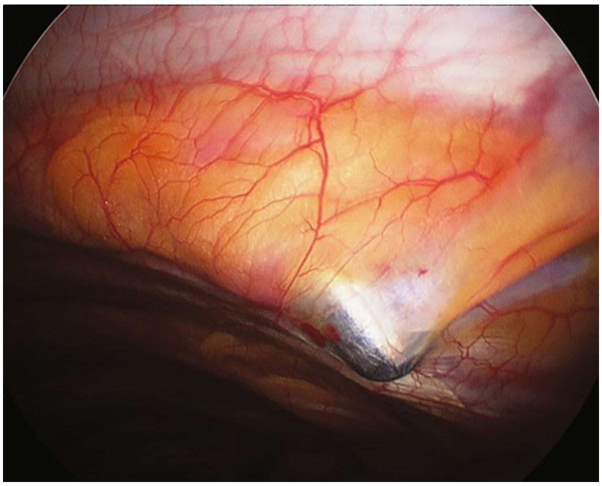

Fig. 3. Endoscopic view of the right pleural space and deflation with blocked right unit of the double-lumen tube. The round dissector appearing at the intercostal space prior to perforation of the parietal pleura

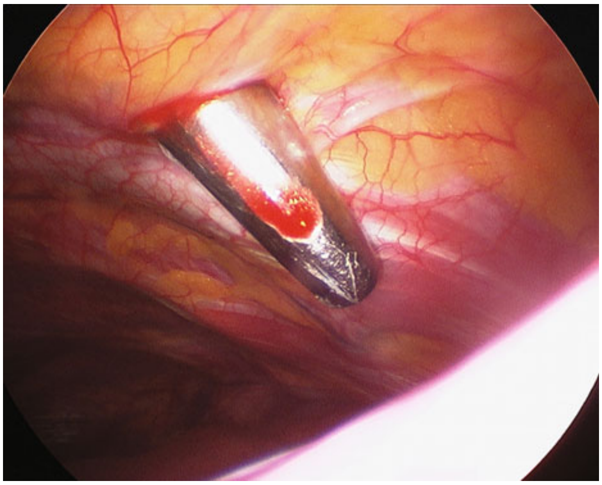

Fig. 4. Same view as in Fig. 3, immediately after perforation of the parietal pleura. Notice almost no bleeding due to the blunt dissection with the aid of the round tunnelizer

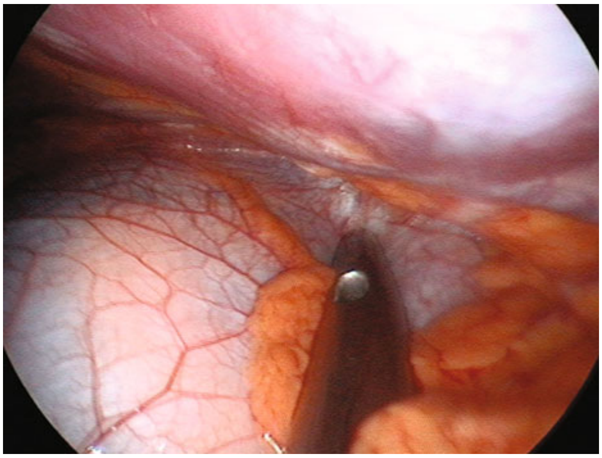

Fig. 5. Blunt preparation of visceral pleura and lung at the right side toward the mediastinal fat and pericardium

especially in adults is mandatory and during setting of the thoracoscopic ports on either side of the respective lung is excluded from ventilation. This setting allows a careful and entire exploration of the respective pleural cavity and blunt dissection with or without electrocautery of eventual adhesions between the visceral and the parietal pleura under controlled intraoperative visualization (Figs. 3–8).

According to our experience, the use of a 5 mm/30° angled optical system is sufficient for a complete exposure of both pleural cavities, also on the left side where occasionally the heart, usually displaced by

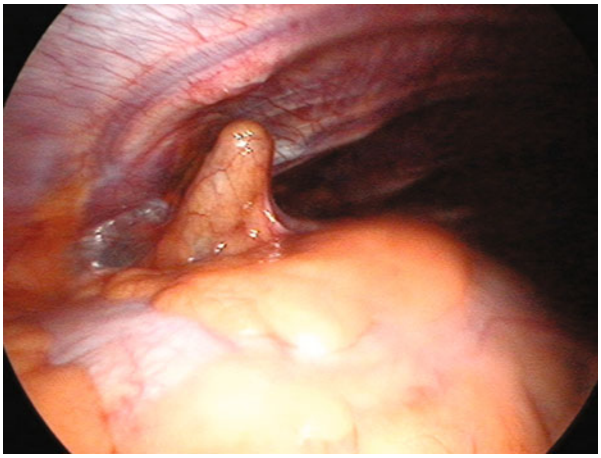

Fig. 6. The round tip of the tunnelizer appears at the left side with some pericardial fat elevated. The endoscope now is introduced at the left side, with the right lung inflated and the left lung deflated

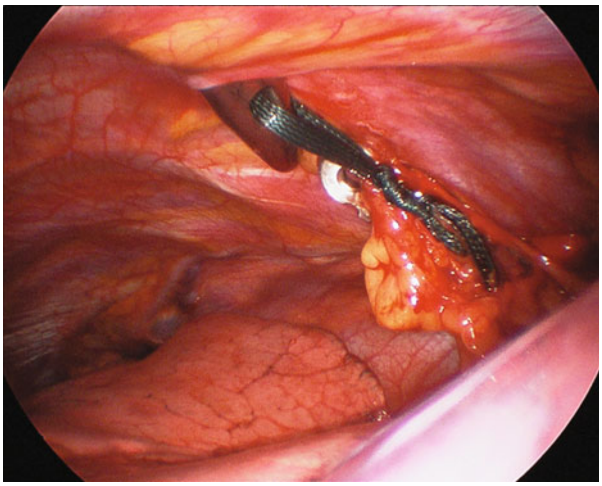

Fig. 7. The situation demonstrates a potential complication with trapped pericardial fat at the pectus bar during implantation and pull through. View at the right pleural space, immediately prior to slipping through the right thoracic wall

the sunken sternum, may render a sufficient overview more difficult. Also, 30° angled optics much easily enable the observation of the thoracic wall from the intrathoracal aspect during insertion (Figs. 4 and 5) and passage of the dissector or the pectus bar, guiding its retrosternal passage maneuver (Fig. 5) between the posterior aspect of the sternum and the heart within the prepericardial fat pad (Fig. 6). In addition, the phrenic nerve may clearly be identified and preserved as the dissector and pectus bar pass ventrally to it.

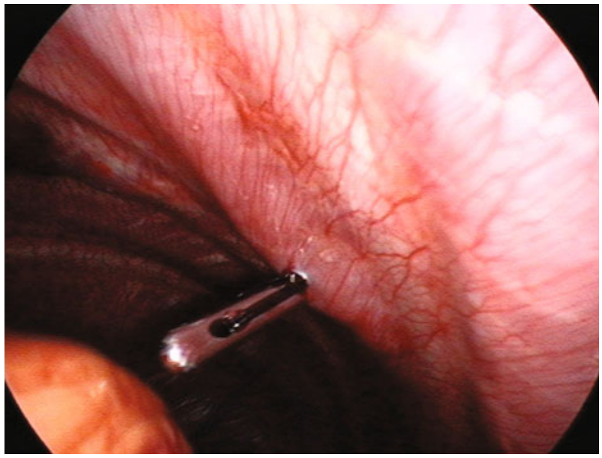

Fig. 8. Endoscopic view at the left lateral thoracic wall, depicting the threaded Deschamps-awl containing a strong suture for circumcostal fixation of the lateral pectus-bar end

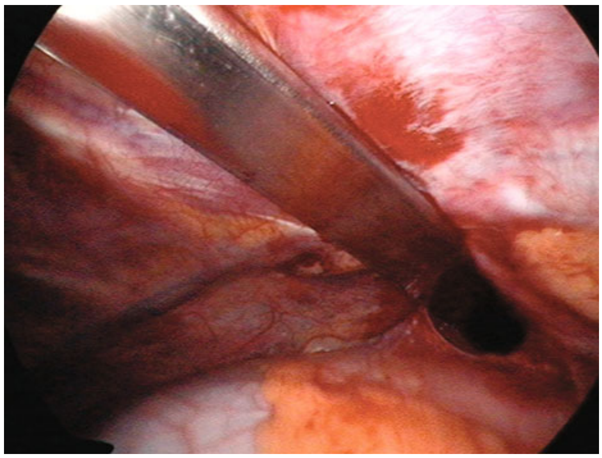

Fig. 9. Right pleural cavity with the pectus bar perfectly in place. The internal thoracic vessels (running from the center of the picture to the bottom left) remain undamaged

Subsequently implantation of the shaped pectus bar firmly attached to the applicator from one to the other side is also performed under thoracoscopic view (Fig. 7), allowing eventual adjustments of bar position during this maneuver and minimalizing any risk for injury to adjacent structures and intrathoracal organs. In the case of stabilization of the pectus bar utilizing circumcostal sutures, the passage of the threaded Deschamps-awl between the ribs may accurately be visualized (Fig. 8) to avoid damage to the lung surface. Finally the correct position of the placed pectus bar is controlled via endoscope (Fig. 9) and potential bleedings or injuries to the intrathoracal organs may be detected in due time.

Additionally and in contrast to 0° non-angled optics, the flexibility of 30° angled systems is superior concerning general intrathoracal overview and an extended survey may be achieved just by simple rotation of the optics along its longitudinal axis into the direction of interest.

Two small skin incisions not extending 5 mm and in accordance with Langer's cutaneous lines are made on either side of the thorax, preferably at the eighth or ninth intercostal space, at the extension line of the anterior axillary fold (Fig. 2). Subcutaneous and muscle planes are divided with small scissors and blunt perforation of the parietal pleura is then obtained, using a small clamp and after alternating exclusion of ventilation on either side in close cooperation with the anesthetist utilizing the technical properties of a double-lumen tracheal respiration tube (Chapter 5.3). Thereafter, a 5 mm trocar–port is inserted through the same incision and screwed into the pleural cavity. Care must be taken to place the incisions and trocars on the superior margin of the rib to avoid subcostal damage to intercostal vessels and/or nerves with potential cumbersome neuralgia. In addition, a symmetrical alignment of both 5 mm cutaneous trocar-port incision sites should be aimed at for aesthetic reasons.

Gas insufflation during videothoracoscopy is not necessary since the pleural space is very well accessible after ventilation exclusion on the respective side, when a double-lumen tracheal tube is used and thus temporary collapse of the right or left lung is obtained. Because the left main bronchus is anatomically longer than the right one, usually a "left-sided" double-lumen tube is used by the anesthesiologist to perform alternating and separated ventilation of right or left lung effectively.

Usually, there is no need for routine insertion of a chest drainage tube ensuing the surgical procedure, except for a potential collection of fluid or blood after placement of the bar(s), or when an air leak at the lung parenchyma is evident. In these cases, a 16 Charriére chest tube generally is sufficient for effective postoperative fluid evacuation and/or management of parenchyma air leakage. It is inserted through the initially performed 5 mm port incision, and fixed to the skin by strong sutures. After radiologic chest control the chest tubes may be removed on the second or third postoperative day depending on the extent of fluid and/or air output through the drainage. The patient is asked to hold breath in expiration and the thoracic drainage is then quickly removed, a swab with disinfectant ointment is subsequently applied at the orifice to avoid air aspiration during drain removal. As an alternative, an air and liquid tight, transparent adhesive tape may be placed instead of the sterile swab, which enables visibility and direct control of the small wounds. The patient should thereafter rest in bed for 1 h with a firmly attached wound dressing. In the asymptomatic patient chest radiographic control is done only on the following day.

As mentioned above, potential complications like intraoperative hemorrhage may be detected early, when VATS is routinely used during the MIRPE. Therefore, a basic thoracotomy emergency set should be available in the theater at any time during surgery, including thoracic rib–retractors of different dimensions depending on patient age, vascular clamps, and lung grasping forceps. Thus, successful and early management of potential intraoperative complications may be achieved, either by continuing in VATS or conversion to open thoracotomy approach if necessary in major problems.

All these aspects have to be considered and discussed with the patient preoperatively in detail, when selection for the pectus-bar implantation is indicated, information about the planned surgical procedure is offered, and signing of informed consent is arranged. Particularly, rare but potential intraoperative hemorrhage due to damage to greater vessels or heart has to be mentioned. Eventual intraoperative surgical complication management, with necessary extension of the minimally invasive access or even conversion to open surgery must be explained to the patient accurately.

5.4.3 Conclusion

Nowadays the routine use of intraoperative videothoracoscopy in the MIRPE has to be strongly recommended in specialized referral centers for surgical treatment of extensive funnel chest deformity. Preferably, and especially in adolescents or adults, the videothoracoscopy should be performed by a thoracic surgeon, who is familiar with the use of such videoendoscopic equipment during standard and complex operations in general thoracic surgery. This surgeon should be able to recognize potential intraoperative complications (Fig. 7) during the pectus-bar implantation, and must be capable and be trained to manage such complications. That is why an interdisciplinary approach is strongly recommended especially in the cases with a higher prevalence to complications, which rather likely occur in elder adolescents and adults [14]. Furthermore this need for interdisciplinary expertise and, in contrast to other routine surgery, the lower number of pectus excavatum cases that are treated, are strong arguments for focussing the selection, indication setting, and performance of surgery as well as after treatment to specialized referral centres, experienced with a sufficient high number of cases treated per year.

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Special techniques in the funnel chest deformity

6

6.1 The Ravitch procedure

Anton H. Schwabegger

With the triumphal march of the surgical method (MIRPE) in the correction of pectus excavatum deformities, according to Donald Nuss [6] the method originally described by Ravitch in 1949 and 1958 [9, 10] was partially dislodged into the background and minor scope was left for special applications only. However the Ravitch technique is still widely used as a standard procedure in the correction of pectus carinatum deformities (Chapter 7.1) Although a recent study consisting of a systematic review and meta-analysis methodology confirmed that the complication rate in the MIRPE technique is higher than in the Ravitch technique, and the period of requirement of postoperative analgesics seems to be lower than in the MIRPE collectives, a clear difference concerning the aesthetic outcome could not be elaborated. It seems that particularly the parameters of pain and aesthetic result, being of paramount importance for the patient self, could not be studied comparatively due to too many biasing factors and lack of long-term comparability [1, 5]. Despite that the MIRPE technique offers a method requiring far shorter surgery time and represents an overall elegant method with however pleasing results lasting for many years. The elegancy and straightforward technique in experienced hands with relatively hidden scars supported its triumphal march so far, convincing patients and surgeons as well. However, these findings are predominantly true for children and adolescents, but must be regarded differentiated in adults. For the latter collective of patients no comparative data are yet available.

The Ravitch procedure is a method to correct either keel or funnel chest deformity by subperichondral resection of the overabundant length of several rib cartilages and thus causes therewith a diminuation of the surface area from a concave funnel or a convex keel into a flat plane. Essential within this method is a horizontal sternum osteotomy in combination with parasternal subperichondral either chondrotomies or chondrectomies at individual levels, depending on the shape of the deformity. While through the implantation of a metal hardware, the so-called pectus bar (Chapters 6.2–6.4) an arching of the funnel deformity into a flat or slightly convex shape is produced and therewith also an advantageous expansion of the thoracic cavity is produced. On the contrary, in the Ravitch method such additional intrathoracic space is accomplished to a minor extent because almost no extension of the thoracic cavity is feasible without such a distension device, which a pectus bar represents. In spite of the nowadays very widely used MIRPE method [6], according to Ravitch, indications still exist for the application of this conventional technique without the use of a metal bar. Certain patients or parents refuse the implantation of metal hardware itself, are alienated by the potential complications of the endoscopically assisted intrathoracic manipulation, or simply show somatic intolerance or known allergic reactions against metal ingredients like the component Nickel.

However, the Ravitch method still suits very well for purely aesthetic corrections of minor funnel deformities (Figs. 1 and 2), in children as in adults, when alternative methods such as custom-made silicone implants (Chapter 6.5) are rejected by the patient or the alternative of lipofilling (Chapter 6.6), based on lacking autologous fat in slim or athletic patients, herein yet in adolescents and adults, is not feasible. A further indication for the utilization of this conventional technique exemplarily is a simultaneous intervention in patients with open heart or vessel surgery simultaneously suffering from a funnel chest deformity (Chapter 6.7). Occasionally and predominantly patients with pectus excavatum prior to all pectus deformities after decades of long course may show unrecognized heart valve problems [2, 3, 8, 12, 13, 16] so that in the midlife adulthood then a heart valve operation becomes necessary. On this occasion also the funnel deformity can be corrected simultaneously with the conventional method [7, 11]. On the other hand, an arching of the funnel deformity with a metal pectus bar placed horizontally at the anterior thoracic wall in these patients operated at the greater vessels or heart represents a contraindication. Because in the eventual case of an acute intrathoracic complication, an immediate surgical intervention

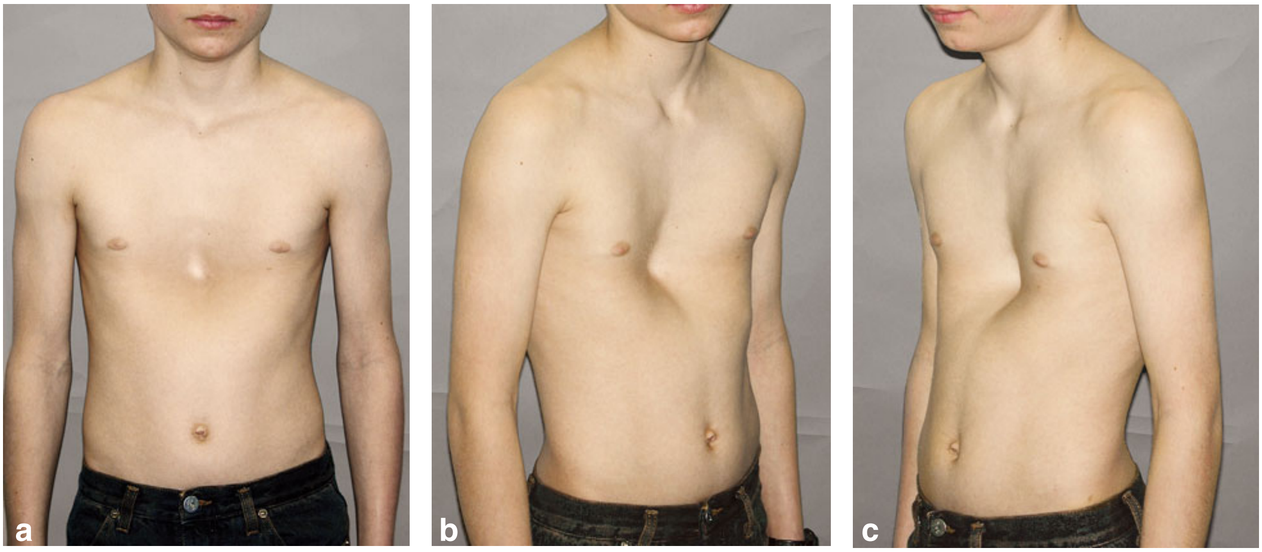

Fig. 1a–c. 14-Year-old male with major pectus excavatum deformity presenting with a steeply plunging funnel causing restriction of cardiopulmonal function. The parents refused invasiveness through MIRPE, thus the conventional Ravitch technique was planned

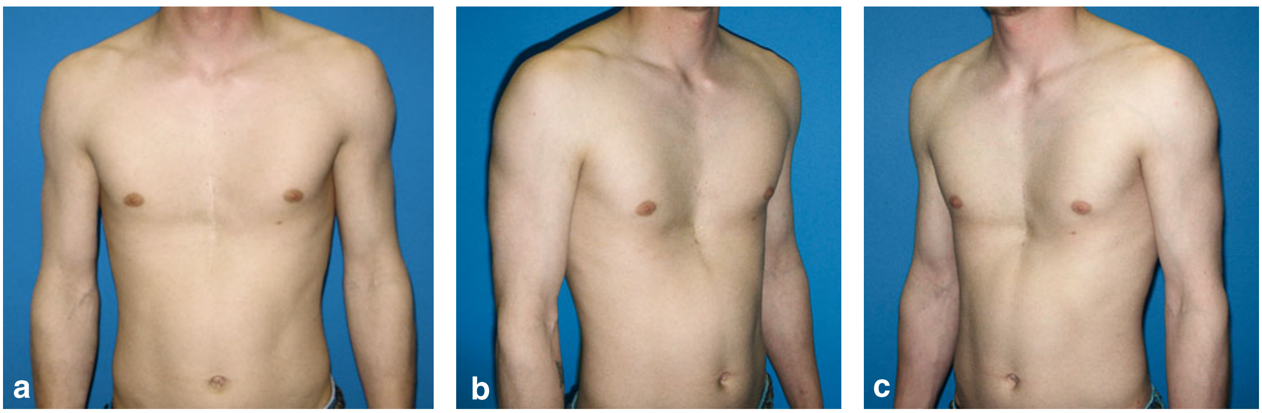

Fig. 2a–c. Same patient 7 years after the Ravitch procedure with stable situation and overall patient satisfaction, but objectively minor relapse. Patient is working as a mason without cardiopulmonal impediments

into or within the thoracic cavity is then blocked by the presence of such metal hardware and may even complicate acutely necessary radiographic or magnetic imaging. Furthermore delivery of cardiopulmonary resuscitation in such patients with a pectus bar in situ seems to be extremely challenging due to its rigidity, however designed to withstand any reflexing forces of the thoracic skeleton [17]. Alternatively absorbable plates and screws (Chapter 6.2) can be used [3, 13, 14] herein, which can be cut or transsected easily in the case of an acute intervention with conventional instruments or do easily give way to resuscitation maneuvers.

6.1.1 Surgical technique of the Ravitch procedure

In male patients, a vertical midsternal skin incision is preferred (Figs. 1–3), but may be modified according to the desires of a patient into an oblique or a horizontal fashion (Chapter 5.1). In female patients, on the other hand the image of the décolleté should remain scar free, thus the surgical access becomes somewhat awkward through modified unilateral or bilateral curved incisions, in order to hide the scars along the submammary crease likewise to the incisions performed in the keel chest deformity (Chapters 5.1, 7.1, 7.2 and 9).

Fig. 3. Extent of number and length of rib cartilage resection in the Ravitch procedure. Also note both the horizontally orientated bony wedge resections at the 3rd and 5th intercostal spaces as well as the resected xiphoid

The dissection of the skin and fat flaps off the musculature succeeds in a usual way. In minor to moderate deformities [15], the subsequently necessary chondrotomies and chondrectomies may be accomplished also by means of the muscle split technique (Chapter 7.2). Each individually disfigured and dissected rib cartilage becomes exposed with a door-like incision at the perichondrium (Fig. 4a). The elevation of the cartilage succeeds with a raspatory (Fig. 4b) while resection is performed using a rongeur. The transsection ideally takes place by means of small scalpel (Fig. 4c) or diathermic cautery. That much of cartilage must be resected and that many incisions at the remaining rib cartilage must be performed that an unbending of the deformed rib portion is enabled without essential tension (Figs. 5 and 6). Prior to any unbending of the deformed anterior thoracic wall, a horizontal osteotomy must be undertaken at the location, where the sternum is inclinated to dorsal. This succeeds with oscillating saws and is carried out solely at the ventral compacta and the Cancellous bone. Such a horizontal osteotomy requires a bony wedge resection (Figs. 5 and 6) in order to allow a forward bending and elevation of the depressed sternum.

At the same time it is to be avoided herewith that also the dorsal compacta will be transsected, because otherwise through dislocation of the mobilized distal sternum part an unsightly and well palpaple ridge may develop, moreover a painful pseudarthrosis might remain. In a

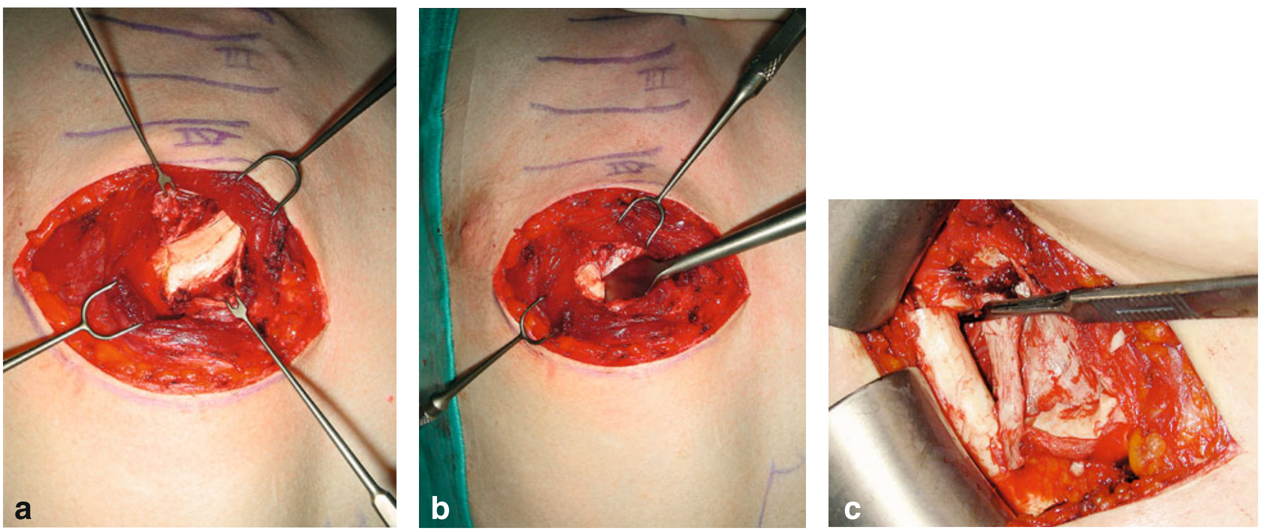

Fig. 4a. Door-like incision and dissection of the perichondrium via the muscle split access to expose the deformed rib cartilages at each individual level. b Dissection of the deformed rib cartilage and elevation out from the perichondrium tube. c Transsection of the rib cartilage by scalpel with particular attention not to damage pleura, internal thoracic vessels, and intercostal nerves

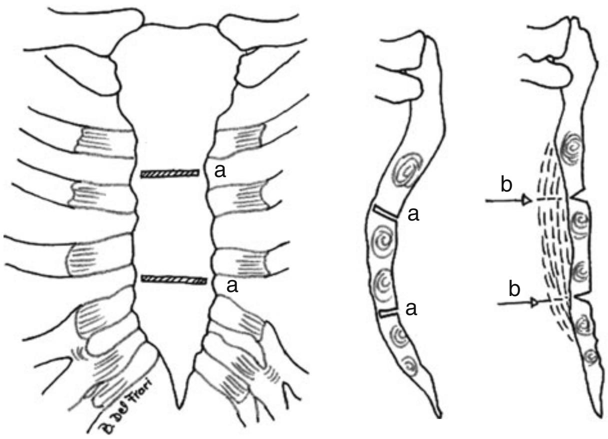

Fig. 5. Schematic depiction of horizontal osteotomies (*a*) at the sternum, the level of singular or multiple incisions may differ depending on the shape of deformity and aimed remodeling. Unbending (*b*) of the incised part of the sternum with green-stick fracture of the dorsal lamina in order to create its final shape

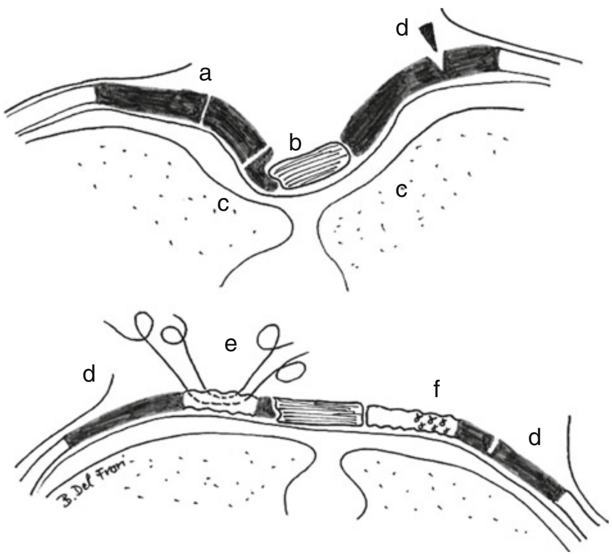

Fig. 6. Deformed cartilages with abundant length (*a*) will be partially resected and for the case of sternum malrotation (*b*) a horizontal osteotomy must be performed. The lungs (*c*) will follow the remodeling procedure. The remaining rib cartilages, if still showing protrusion, need wedge resections at single or multiple sites (*d*) to allow unbending and giving way to the shortening effects on the perichondrium tubes by reefing sutures (*e*, *f*)

female, for the case that the surgical incision was selected to be placed at the submammary crease, the access to a far cranially sternum osteotomy is impeded by an oblonged subcutaneous tunnel. Herewith the utilization of angled oscillating saws is strongly recommended (Fig. 16 in Chapter 7.2) in order to avoid undue skin overexpansion or malplacement of the osteotomy itself. After osteotomy and multiple chondrotomies and/or chondrectomies, the elevation of the now loosened sternum follows by digital mobilization and separation from the mediastinal organs. Subsequently the elevation of the sternum into the horizontal plane is managed for instance with a bone hook and is held in this position now. Simultaneously with unbending the inclination of the sternum, the dorsal, however not transsected compacta, breaks similar to a green-stick fracture but must remain without any dislocation. In contrast to the limited incision at the bone, the dorsal compacta may or in distinct cases even has to be transsected, particularly in cases when malrotation or major twisting of the sternums exists along the longitudinal axis or deviation out of it to lateral is present. Especially in these cases where the dorsal compacta is subject of total transsection, the use of osteosynthetic material, either wires, strong sutures, or plates (Chapter 6.2) is unconditionally necessary for the prevention of relapse of the deformity or unintentional malrotation through muscle tension. In such cases with major deformities and sternum malrotation, a three dimensional CT-scan, utilizing the volume rendering mode in order to accurately depict especially the cartilagineous structures is mandatory [14]. Such an imaging will alleviate the preoperative planning of the extent and site of chondrectomies, chondrotomies, and finally the osteotomies at the sternum (Fig. 7 in Chapter 3.3). The perichondrium tubes are then shortened with heavy absorbable PDS suture, the so-called reefing sutures (Fig. 5).

For the case that by these reefing sutures the elevated sternum cannot be held in the desired position, it can additionally be mounted with the aid of absorbable plates or mesh and screws (Chapter 6.2) in the zone of the sternum osteotomy.

After completion of the remodeling process the muscles are sutured or refixed in anatomically orthotopic position.

The elevation and an even intended slight arching forward of the anterior thoracic wall using the Ravitch procedure especially in adolescents and adults hardly (Fig. 7) succeed to the same extent as such is feasible with the suspension of pectus bars, either applied endoscopically or with an open access. Therefore the patient prior to selecting an appropriate method and prior to planning a surgery has to be informed on the circumstances of limited extent of remodeling capacity using the conventional Ravitch procedure, particularly

Fig. 7. 27-Year-old male, corrected with the conventional Ravitch technique, 1 year after surgical correction of a pectus excavatum deformity, resulting in major improvemet of the deformity. The still present depression may be due to lacking internal suspension in already matured and rigid skeletal structures

in adults. However a distinct improvement of a funnel deformity may yet be achieved but with probably higher incidence of at least partial relapses by time, in contrast to and according to the actual experiences with the methods using metal hardware for sternum suspension.

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6.2 Modifications of the Ravitch procedure and similar methods

Anton H. Schwabegger

There is no ideal method for all seasons.
Ancient medical wisdom

Several modifications of the conventional however genial Ravitch procedure have been developed over the last decade, all of them intending to support the elevated sternum with a diversity of materials. The conventional technique is based on rigid remodeling utilizing reefing sutures to produce a jumping sheet-like flat anterior thoracic wall. The tight reefing sutures aim at long-lasting tension and scarring with a permanent result. Such intervention may be feasible in children with plasticity of immature thoracic skeletal structures, a rapid healing process, and almost neglectable biomechanical strains from the musculature. In contrast to that in adults such reefing sutures, even with the aid of sternal bone relaxation by osteotmies, will hardly result in sufficient sternum elevation and moreover the long-term result is endangered by the rigidity of the adjacent skeletal components, the biomechanical memory of artificially bended cartilage and the muscular strength. In order to support any remodeling in adolescents and adults, metal wires or struts have been advocated since the mid of the last century [1, 3, 4, 7–9, 11, 16, 18, 20, 21, 23, 24] and have found continuation in the finally refined MIRPE technique, ultimately avoiding major surgical invasiveness of a remodeling procedure to the anterior thoracic wall. Bioabsorbable sheets and plates [5, 13, 14] or non-absorbable Marlex® [17] or similar alloplastic material shaped as mesh, threads, or straps may also serve to support the sternum and to prevent re-depression. Even autologous materials like rib struts [6, 15] or fascias to be placed underneath an elevated sternum have been used. However, also such autologous tissue may be well utilized in children with sufficiently pleasing results, but will be suitable not at all for an appropriate correction in adults because of the above-mentioned reasons.

The Hegemann procedure [8], performing a combined approach finally gave way to a nowadays-established technique worldwide, a method ideally suitable for many types of deformities, but gradually and to a major extent replaced by the MIRPE technique already. This

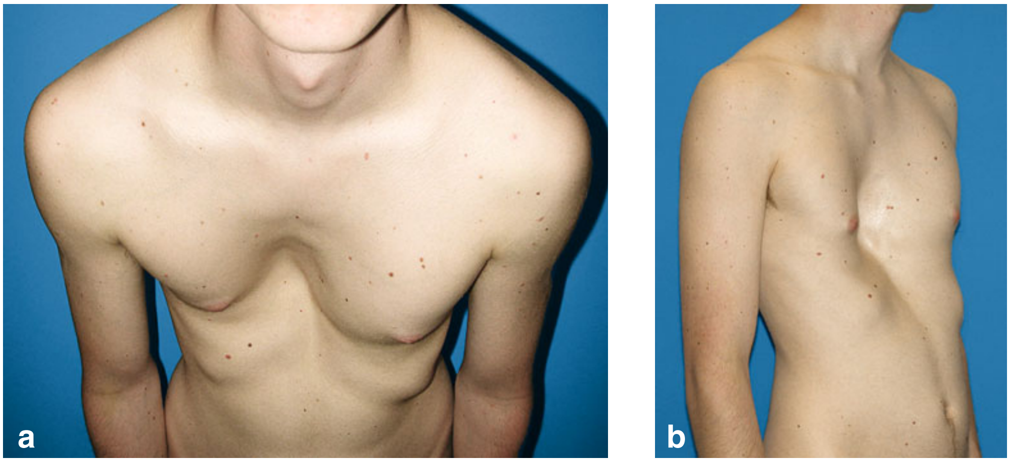

Fig. 1a and b. 20-Year-old male with severe asymmetric pectus excavatum and manifest Nickel allergy, inaccessible for the utilization of metal hardware to correct the deformity

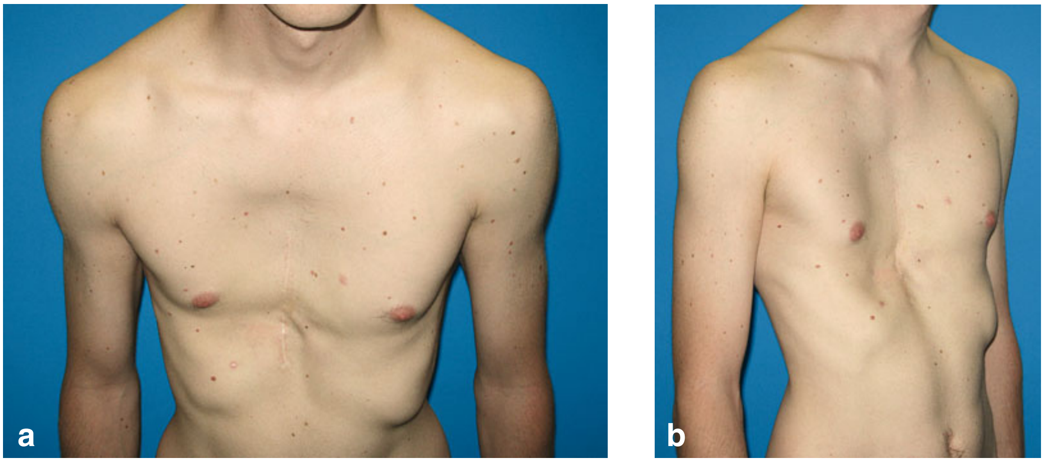

Fig. 2a and b. Same patient with moderate aesthetic result, 1 year after modified Ravitch repair, using a bioabsorbable mesh plate to fix the derotation of the elevated sternum

ingenious combined technique, described and advocated by Hegemann in 1965 [8] and Willital in 1981 [24], relies basically on the Ravitch technique, but complemented with a horizontally placed short metal strut, implanted transsternally with lateral rest on the ribs, which form the funnel margin. Other authors were more invasive in utilizing even two further parasternal metal struts to fix the multiply chondrotomized rib cartilages into a frontal plane [20].

Within all of these variants, the sublay of material of large surfaces, that need sternum and attached ribs to be separated from each other, should be avoided. Such a wide ranging detachment leads to disruption of the terminal branches of the intercostal nerves from the end organ, the parasternal intercostal muscles and periosteum, and overlying skin (anterior cutaneous branch of the intercostal nerve). Lesser the resulting numbness or lowered function of the affected intercostal muscles annoys the patient, but the more will potentially growing neuromas after years lead to cumbersome disturbing pains with abuse of analgetics [19].

A large number of publications so far, impossible to be all cited here, deal with minor modifications of both these basic concepts, namely the loosening of the rigidity of the deformed anterior thoracic wall with multiple incisions of cartilages and bone, and furthermore the support of the sternum in order to prevent relapse. Any minor modification should be considered as far as it is known, but must be adapted to the individual requirements of patients and the remodeling procedure, however following the basic principles when alternative and more modern methods are refused or excluded by other reasons.

6.2.1 Surgical technique

Out of a variety of modifications exemplarily a technique with absorbable osteosynthetic material is presented here (Figs. 1 and 2). The surgical approach generally does not

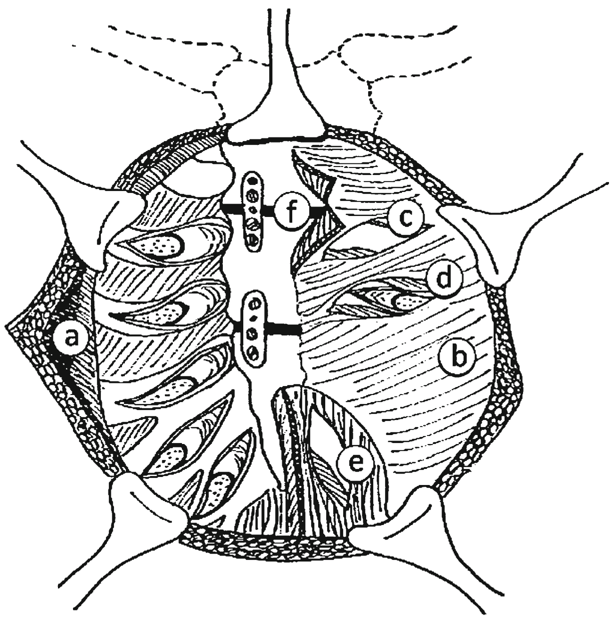

Fig. 3. Schematic overview of conventional technique at the patient's left side and muscle split technique at the right side. (*a*) Retracted pectoralis major muscle and skin flap with rib cartilages resected (*b*). Pectoralis major muscle left in place with several transmuscular splittings (*c*) and (*d*, *e*) muscle split along the rectus abdominis muscle to expose the 7th and 8th rib cartilages, (*f*) slightly elevated periosteum and horizontal sternum osteotomy with absorbable osteosynthetic material

differ from that described already in the previous chapter (Chapter 6.1). Also and preferably the muscle split technique [22] is applied to access the affected ribs, when feasible accordingly to the extent of the deformity. The periosteum is elevated and the sternum is incised at the region of the sternum inclination and if necessary also in a potentially present ascending lower part (Fig. 5 in Chapter 6.1). Prior to the application of the reefing sutures (Fig. 6 in Chapter 6.1), the osteotomized sternum is elevated into a frontal plane either by blunt separation from the intrathoracal organs using the finger, or it may be elevated using a bone hook. Resting in the desired position, the reefing sutures are set. Without relaxation now the osteosynthetic absorbable material is fixed to bridge the osteotomy incisions. For the case, that a bony wedge is resected at the zone of inclination, this wedge may be reimplantated at the lower incision, which is subject of creating a gap due to the unbending maneuver. That piece of bone is then attached and kept in place by the plates (Figs. 3 and 4; Fig. 10 in Chapter 7.2). Particularly in cases with sternum malrotation, when a complete horizontal osteotomy, which includes transsection of the dorsal sternum lamina also is required, the application of an absorbable mesh plate with greater surface is strongly recommended. Especially heat-malleable (PLA/PGA) polymerized glucose-lactide plates (Lactosorb®, W. Lorenz Surgical, FL, USA) can perfectly be adapted to bony incongruences that inevitably result from the derotation maneuver. The greater surface of such a mesh (Fig. 5) contains many more drilling holes than the longitudinal plates and therefore may guarantee much higher stability of the osteotomized and refixed sternum parts for several months and thus until healing of the whole remodeled area [5, 12]. The periosteum is then reattached covering the zone of osteosynthesis and the split muscles are joned with loose adapting sutures. For the case of performed muscle flap elevation to expose a wider area of remodeling, these muscle flaps are united in the midline to cover the whole presternal area, which on the one hand leads to an augmentation of soft tissue volume there in the zone of interest and on the other hand supports rapid healing through the coverage with well-vascularized tissue [18].

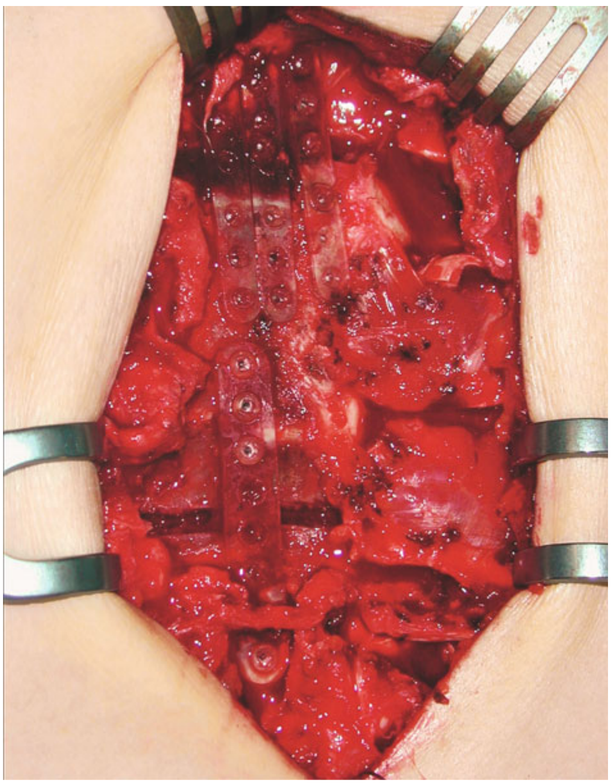

Fig. 4. Intraoperative situs of a modified Ravitch repair of adepressed sternum. Two horizontal wedge resections at the sternum after sternum elevation to a frontal plane are refixed with bioabsorbable plates and screws

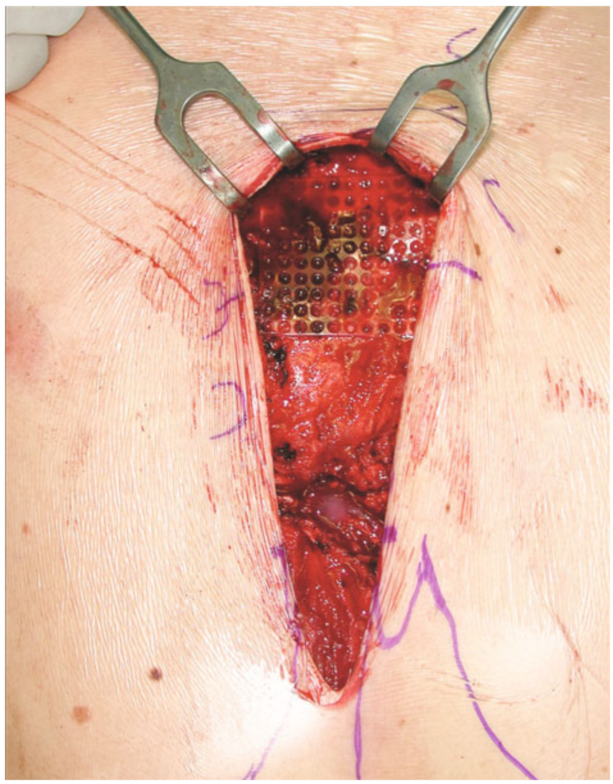

Fig. 5. Intraoperative situs after derotation of a twisted sternum. The fixation is managed with a bioabsorbable mesh, enabling a superior rigid osteosynthesis

6.2.2 Conclusion

A large variety of modifications of the originally described Ravitch procedure exist to correct a funnel chest deformity. Among them the variants that support the position of an elevated sternum with alloplastic material seem advantageous over the conventional method in the generation of permanently pleasing results and in the prevention of relapse, particularly in the collective of patients beyond childhood. No clear evidence but only singular publications [2] in the comparison of different methods are available so far, either due to the diversity of deformities within single institutions or because of different measurement tools worldwide in the evaluation of the outcome of patient satisfaction and objective aesthetic results. The complication rate in utilizing non absorbable materials, in particular with metal struts seems to be higher due to potential migration and dislodgement [13]. This phenomenon is not present with the use of bioabsorbable material, although not providing equal stability than metal devices, they become disintegrated after 1 year and finally will be absorbed completely [10, 12] without producing any known complications so far. However, the choice of an appropriate method should be left to the skilled and well-informed treating specialist, whose decision is based on experience, surgical preferences, available technical equipment, and informed consent with the patient and his/her desires.

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6.3 Thoracic wall deformities

Ann M. Kuhn, Donald Nuss

6.3.1 Introduction

The open technique or Ravitch procedure for the repair of pectus excavatum involved radical resection of cartilages and "short ligaments" and was the procedure of choice until Pena in 1990 and Haller in 1996 [1, 2] drew attention to development of acquired asphyxiating chondrodystophy as a result of this extensive and early resection of the anterior chest wall structures. This was followed by the introduction of the minimally invasive bracing procedure by Nuss et al. in 1997 [3]. The technique has been compared to placing braces on the teeth. It requires no rib, cartilage, or sternal resection and consists of placing a curved steel bar under the sternum using thoracoscopy for guidance [3–5]. The procedure has since gained acceptance by the surgical community [6–10] because of good to excellent long-term results in over 95% of patients.

6.3.1.1 Indications for surgery

During the latter half of the twentieth century it was common practice for surgeons to operate on very young pectus excavatum patients based simply on their clinical evaluation [11, 12]. Since publications by Haller et al. and Martinez et al. drew attention to the risk of acquired asphyxiating chondrodystrophy [1, 2], most surgeons who still use the open technique wait until after puberty to perform the operations. Although the minimally invasive procedure can be performed at any age without risk of chondrodystrophy, the optimum age is just before puberty since the chest is still soft and malleable, the patients show quick recovery, the results are excellent and the risk of recurrence is very low. Patients who have their repair at a younger age and who have their bar removed before puberty are at a slight increased risk of recurrence.

The decision for surgical repair of a pectus excavatum not only relies on the history and physical exam but also strict objective criteria based on the results of the chest CT, pulmonary function tests and cardiac evaluation, including electrocardiogram and cardiac echocardiogram (Fig. 1). Anterior chest wall malformations can range from mild to severe and at our institution less than half the patients are severe enough to warrant surgical correction. Cardiac and/or pulmonary compression causes symptoms of dyspnea on exertion, chest pain with or without exertion and lack of endurance. Pul-

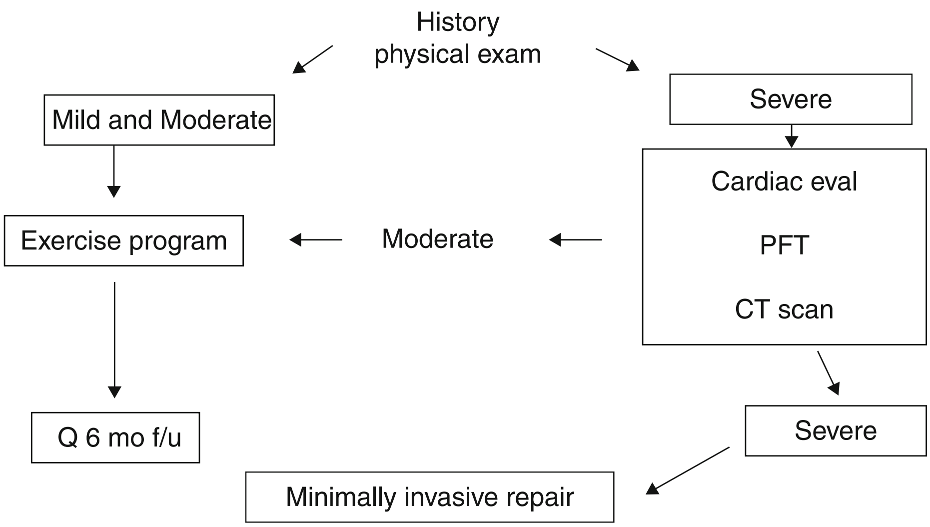

Fig. 1. Pathway for evaluation and treatment of pectus excavatum using the minimally invasive technique

monary function tests often show a restrictive or obstructive deficit. In addition to physical symptoms, a severe deformity may result in poor body image that impacts self-worth [13]. Patients undergoing rapid growth need to be re-evaluated at regular intervals since the deformities tend to progress as the patients grow. This can be especially noticeable during the pubertal growth spurt when a pectus excavatum can progress from mild to severe in as little as 6–12 months.

Classification and treatment algorithm

Initial evaluation of the patient requires a complete history and physical examination including photographs to document the pectus excavatum. Patients are then classified into mild, moderate, and severe categories. The patients with a mild or moderate deformity who are asymptomatic are started on an exercise and posture program and re-evaluated at 12-month intervals. The exercise program is initiated to improve cardio-pulmonary function, chest expansion and to strengthen the chest and back muscles in order to halt the progression of the deformity. Approximately 66% of the patients from our community are treated conservatively. The patients with a severe deformity and patients who have documented progression of their deformity undergo a workup to see whether they have objective evidence to support the need for surgical repair. The workup includes pulmonary function studies, a cardiology evaluation by a cardiologist, including an electrocardiogram and echocardiogram, and finally, a chest CT scan. For consistency, radiologists have suggested that the chest CT be performed during quiet respiration, not during maximum inspiration (Fig. 2). The need for surgical correction is determined if the patient has two or more of the following criteria: (1) a Haller index of greater than 3.25 demonstrated by CT scan; (2) cardiology evaluation demonstrating cardiac compression or displacement (Fig. 3) mitral valve prolapse, murmurs or conduction abnormalities; (3) pulmonary function studies showing restrictive and/or obstructive lung disease; (4) progression of the deformity with associated physical symptoms other than isolated concerns of body image; (5) recurrent pectus excavatum after a failed previous repair by a Ravitch procedure or a minimally invasive procedure.

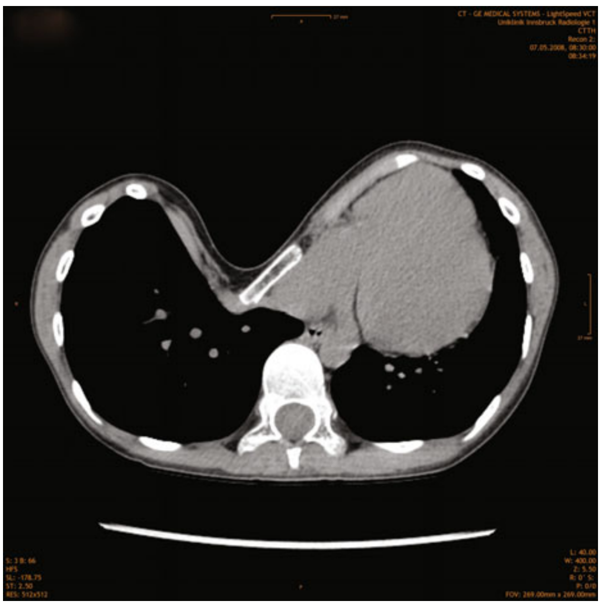

Fig. 2. CT scan showing severe pectus excavatum performed during quiet respiration

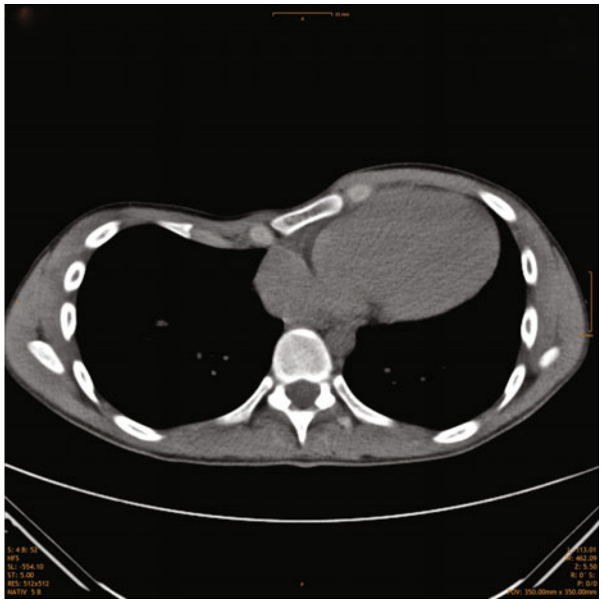

Fig. 3. CT scan demonstrating cardiac compression and displacement, as well as pulmonary compression

Preoperative assessment should also include asking about a history of metal allergy in the patient and immediate family. Nickel and Cobalt are components of the surgical steel used in the bar and are the cause of allergy in 2% of patients [14]. If a metal allergy is suspected, testing by a T.R.U.E. patch (Allerderm, Phoenix, AZ) can clarify this issue. In the presence of metal allergy, a non-allergic Titanium bar is used instead. Unlike the stainless steel bar, which is bent by the surgeon at the time of operation, titanium, must be bent in advance at the factory using CAD/CAM technology.

6.3.1.2 Exercise program

Most patients with pectus excavatum lead a very sedentary lifestyle and have a classic "pectus posture" which aggravates the deformity and may lead to progression of the deformity. Patients who are diagnosed as having a mild or moderate deformity are started on an exercise and posture program designed to improve cardio-pulmonary function and improve posture. Exercise can also increase chest expansion. Breathing and posture exercises are taught to the patients and they are instructed to do these on a daily basis. Patients are also encouraged to participate in aerobic activities such as team sports that involve swimming, running, etc.
Patients are re-evaluated at 6- to 12-month intervals to monitor compliance with the exercise program and to check progression of the deformity. It is possible to halt the progression in a mild deformity and slow the progression in a moderate deformity.

6.3.2 Surgical repair

Although the minimally invasive procedure can be performed at any age, the optimum age is just before puberty. The chest is still soft and malleable before puberty and the patients show quick recovery, a rapid return to normal activity, and have excellent results. The surgical correction of the pectus excavatum is accomplished by inserting a convex steel bar under the sternum with the convexity facing posteriorly. When the bar is in position, it is turned over 180° thereby correcting the deformity. The technique is made possible by the malleability and flexibility of the anterior chest wall and requires no cartilage incision or resection and no sternal osteotomy. After puberty the flexibility of the chest wall decreases making the repair more difficult and patients frequently require the placement of two bars. At our institution we have performed the procedure up to age 31 with equally good results [4]. Other authors have obtained excellent results in adult patients up to 50 years of age [15, 16].

6.3.3 Surgical technique

The preoperative checklist on the day before surgery includes reviewing all studies, checking for allergies including allergy to nickel, and measuring the chest to determine bar length. A measurement from the right to left mid-axillary line is taken and then 2 cm (±1 in.) is

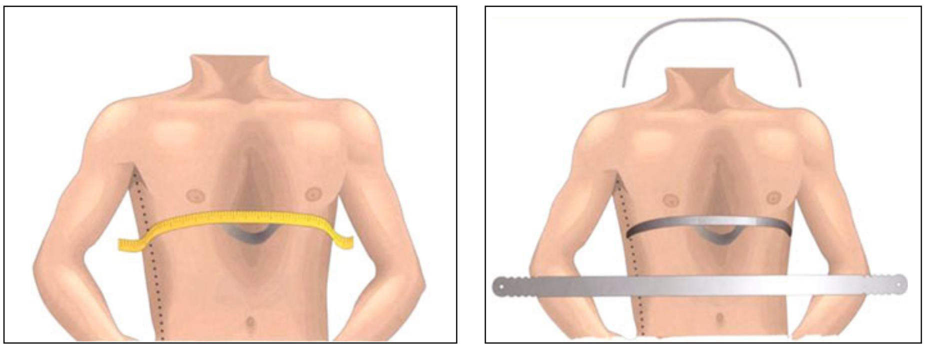

Fig. 4. Technique of measurement determining bar length. The bar needs to be 2 cm shorter than the measurement

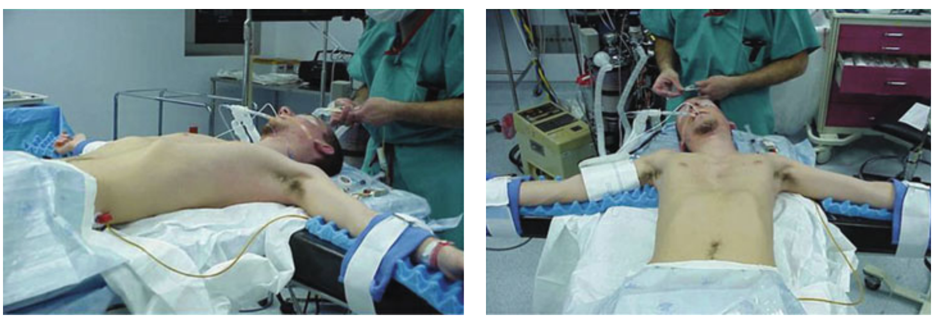

Fig. 5. Positioning of the patient to minimize brachial plexus injury

subtracted from this measurement. The bar takes a shorter course than the tape measure and consequently needs to be 2 cm (±1 in.) shorter than the measurement (Fig. 4).

General endotracheal anesthesia and epidural catheter insertion is undertaken by the anesthesiologist. The epidural catheter infusion is continued for 3–5 days with the average at our institution being 3 days. An indwelling bladder catheter is placed and this remains until postoperative day 1 at which time it is removed. Antibiotic coverage is provided and continued to discharge to minimize the risk of pneumonia with subsequent bar infection. Both arms are abducted at the shoulder and gel pads are used to prevent brachial plexus injury (Fig. 5). There should be slight flexion at the elbows. The patient is draped and the Lorenz™ pectus support bar is bent into a semi-circle, leaving the central 2 cm flat to support the sternum (Fig. 6). Bending the bar into an arch shape allows sustained load bearing of the bar. If the central flat section of the bar is too long, there will be undercorrection of the pectus excavatum.

Fig. 6. Demonstration of a properly bent pectus support bar. The middle bar is correctly bent. The upper bar is too flat, while the lower bar is too rounded

Marking the patient requires determining the deepest point of the pectus excavatum and marking this area with a surgical marking pen (Fig. 7). If the deepest point of the pectus is inferior to the sternum, then the inferior end of the sternum is marked instead. This point sets the horizontal plane for bar insertion. The intercostal spaces that are in the same horizontal plane as the deepest point of the pectus excavatum are marked with an "×". These thoracic entry and exit points on each side of the sternum should be medial to the top of the pectus (costochondral) ridge. Lines are drawn for the proposed incision sites on each lateral chest wall in the same horizontal plane (Fig. 7).

A thoracoscope is inserted through the right lower lateral chest wall approximately two interspaces inferior to the proposed right lateral thoracic wall skin incision (Fig. 8). A thorough inspection of the right hemithorax and mediastinum is performed checking that there is no contraindication for repair. Then pressure is applied to the intercostal spaces marked for bar insertion to ensure that the external markings line up well with the internal anatomy. After confirming by thoracoscopy that the internal and external anatomy match up well, bilateral thoracic skin incisions are made in the region of the mid-axillary line and a skin tunnel is raised anteriorly toward the intercostal space marked with an "×", medial to the top of the pectus ridge (Fig. 9). Also, a pocket is created for the distal end of the pectus bar and stabilizer. Under thoracoscopic

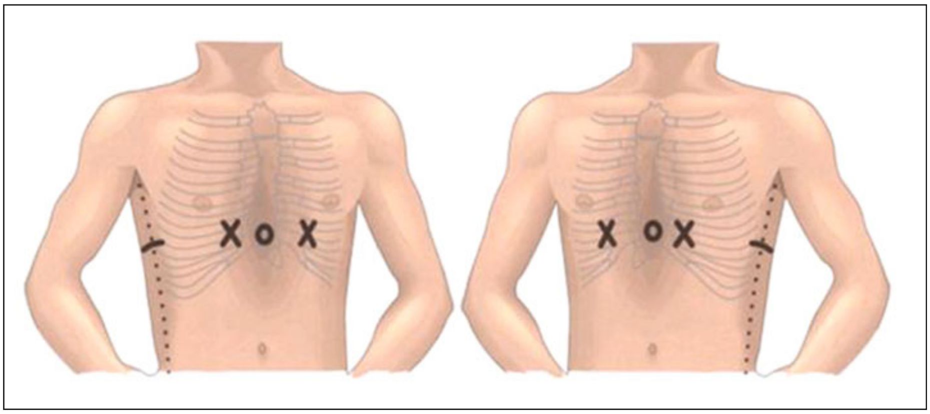

Fig. 7. Marking the bar insertion site. The skin incision, entry and exit sites and the deepest point of excavatum are all in the same horizontal plane

Fig. 8. Placement of thoracoscope for minimally invasive repair

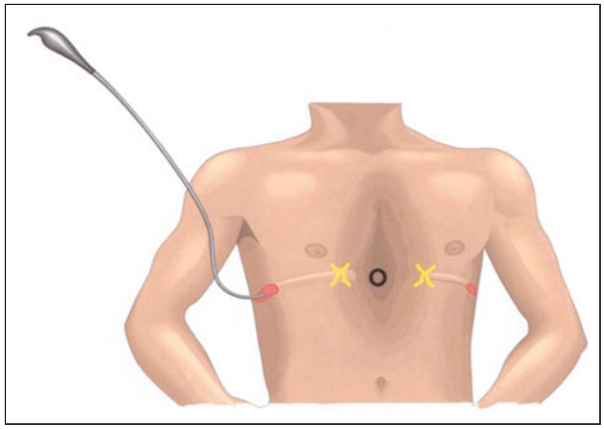

Fig. 10. Insertion of the introducer

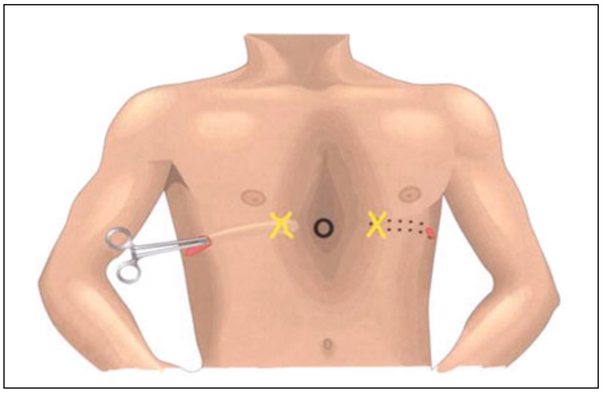

Fig. 9. Creation of the skin tunnel

control, the appropriate size Lorenz™ introducer (Biomet Microfixation, Jacksonville, FL) is inserted through the right intercostal space at the top of the pectus ridge at the previously marked "×" (Fig. 10). The EKG volume is turned up so that the heartbeat is clearly audible. The pericardium is gently dissected off the under surface of the sternum (Fig. 11). The introducer is *slowly* advanced across the mediastinum under thoracoscopic guidance with the point always facing anteriorly and in contact with the sternum. When the substernal tunnel has been completed, the tip of the introducer is pushed through the contralateral intercostal space at the previously marked "×", and advanced out of the skin incision (Fig. 12).

The introducer is then used to elevate the sternum. The surgeon lifts the introducer on the right side and the assistant lifts on the left side (Fig. 12). The lifting is repeated until the sternum has been elevated out of its depressed position and the pectus excavatum has been corrected. An umbilical tape is attached to the introducer, which is slowly extracted, pulling the umbilical tape through the substernal tunnel (Fig. 13). The previously prepared pectus bar is tied to the umbilical tape and guided through the substernal tunnel using the umbilical tape for traction and the thoracoscope for vision. The bar is inserted with the convexity facing posteriorly. When the bar is in position it is rotated 180° using the bar flipper (Fig. 14).

If the bar requires further bending, it is turned over and molded where required using the small Lorenz bar bender. If one bar is not enough, then a second bar is inserted

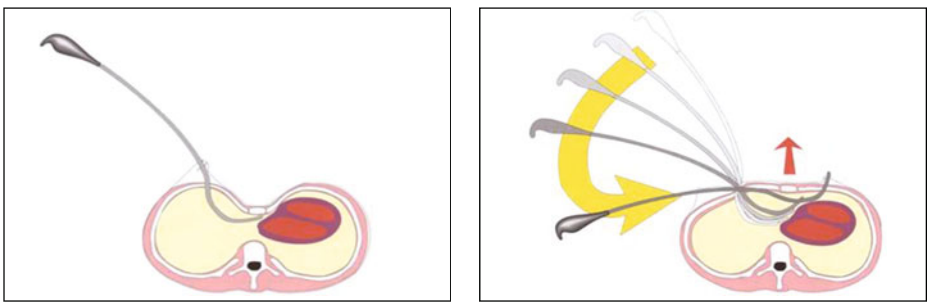

Fig. 11. Technique of dissection with the introducer

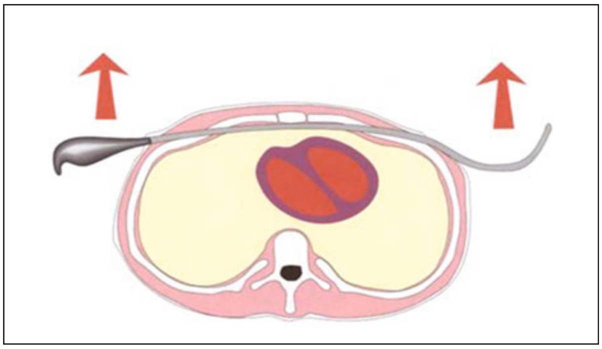

Fig. 12. Demonstration of placement of introducer and subsequent lifting on the sternum

one interspace above or below the first one. Two bars give better and more stable correction, especially in older patients. Slight overcorrection is necessary to prevent recurrence after the bar is removed. Some surgeons prefer approaching the mediastinal tunneling from the left rather than the right side. Other surgeons make a third incision over the xiphoid and then apply a towel clip to elevate the sternum and others do a finger dissection under the sternum before inserting the introducer [17].

6.3.3.1 Stabilization of the bar is absolutely essential for success

A stabilizer is inserted onto the left end of the bar and wired to the bar with No. 3 surgical steel wire. If the bar does not seem stable, a second bar rather than a second stabilizer is probably required. Heavy absorbable pericostal sutures of "0" or "1" PDS are placed around the bar and underlying rib using an "endo-close" laparoscopic needle under thoracoscopic vision on the right side (Fig. 15). Hebra and Gauderer advocated placing a suture adjacent to the sternum [18]. Once the bar is stabilized, the incisions are closed in layers and the pneumothorax is evacuated using the trocar and attached tubing (or a chest tube), with a "water-seal" system. A chest X-ray is obtained before the patient is taken out of the operating room, to check for a residual pneumothorax.

6.3.4 Postoperative management

In the recovery room the patient is kept well sedated with the goal of a smooth emergence from anesthesia.

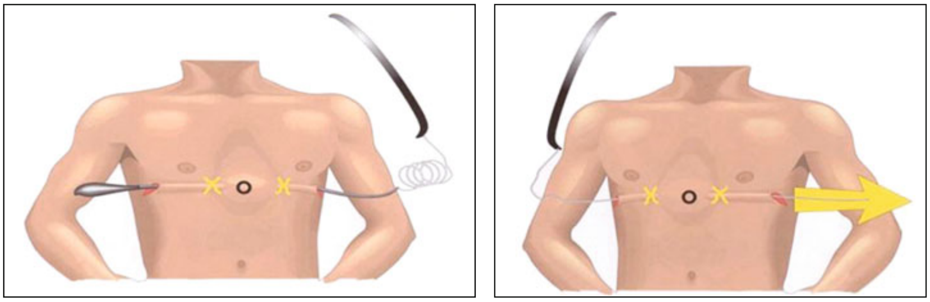

Fig. 13. Technique of using umbilical tape to guide the pectus support bar through the substernal tunnel

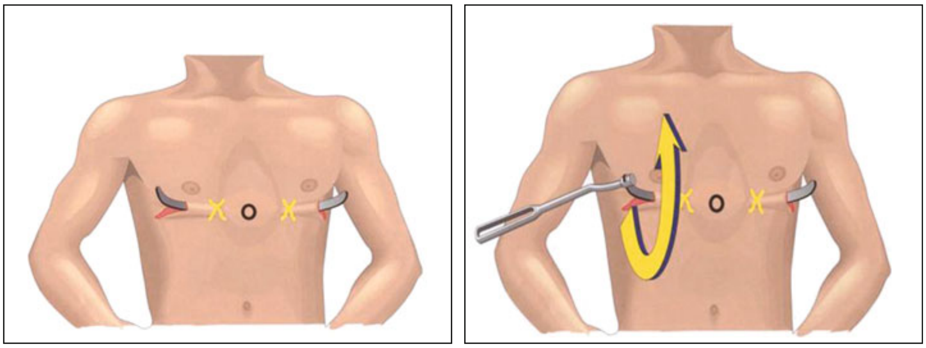

Fig. 14. Rotation of the pectus support bar using the bar flipper. Rotation may be clockwise or counter clockwise

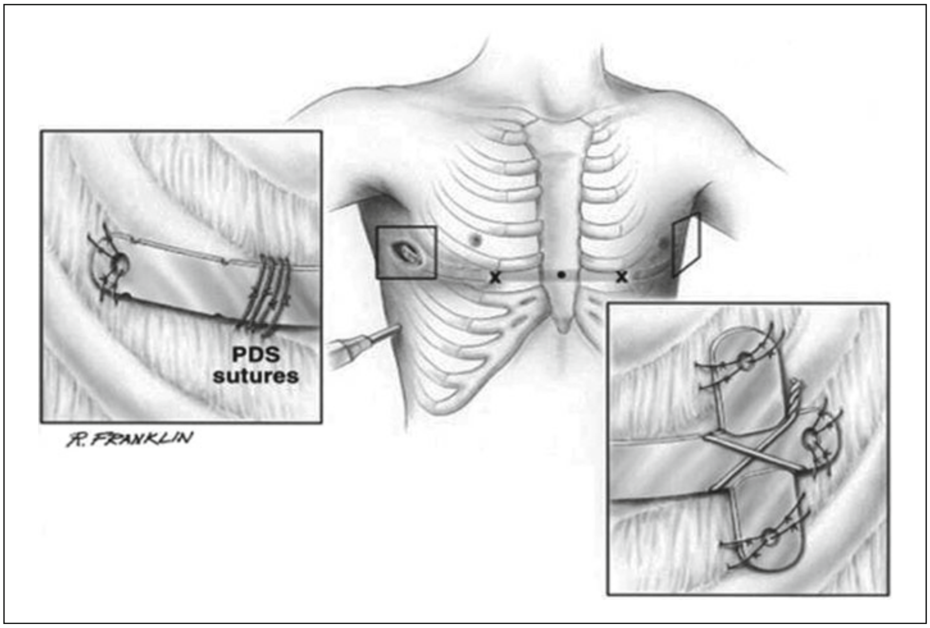

Fig. 15. Stabilization of the pectus support bar

The epidural catheter is left in for 3–4 days. The patient is discharged home on the fourth or fifth postoperative day. Patients may return to school after 2 weeks, but may not participate in sports for 6 weeks from the time of surgery. After the 6 weeks the patients are encouraged to resume their pectus breathing and posture exercises and to participate in aerobic sporting activities (soccer, basketball, and swimming). Heavy contact sports (boxing, American football, and ice-hockey) are prohibited until bar removal.

6.3.5 Technique of bar removal (2–4 years after insertion)

The patient undergoes general endotracheal anesthesia with 5–6 cm of PEEP. The patient is positioned supine with both arms abducted at the shoulder. The chest X-rays are reviewed to confirm the position of the stabilizers. Palpation is then performed to see if bar and stabilizer(s) are palpable and close to the old scar. If the bars and stabilizer are not palpable, then "C-arm" fluoroscopy can be used to determine exact site of the hardware. Use the old scars for incision site if at all possible when removing the bar and stabilizer. The bar ends and stabilizers are mobilized and the wire is cut in two places and removed.

When bar and stabilizer have been freed up from the surrounding scar tissue, the inferior wing of the stabilizer is delivered out of the incision followed by the end of the bar and finally the superior wing of the stabilizer. The stabilizer is removed from the bar. The bar is unbent with the bar flippers or multibenders. An orthopedic bone hook is then passed through the hole in the end of the bar and gentle traction is used to slowly extract the bar. The patient is kept on PEEP until the incision is closed.

6.3.5.1 Timing of bar removal

We advise that the pectus bar be left in place for 2–4 years with most patients having their bar(s) out at 3 years. Patients are evaluated on an annual basis and their growth and activity level are monitored. They are encouraged to do their pectus exercises and to participate in aerobic sports. Patients between the ages of 6 and 10 years and 18 years and older often do not grow rapidly, and they tolerate the bar well for up to 4 years. On the other hand, teenagers who undergo a massive growth spurt (15 cm) may require bar removal after 2 years.

6.3.6 Results

6.3.6.1 Demographics

As of December 31, 2007 we have evaluated 1,941 patients with chest wall deformity. The 1,101 patients have undergone pectus repair, 1,015 have undergone primary operations and 86 have undergone redo opera-

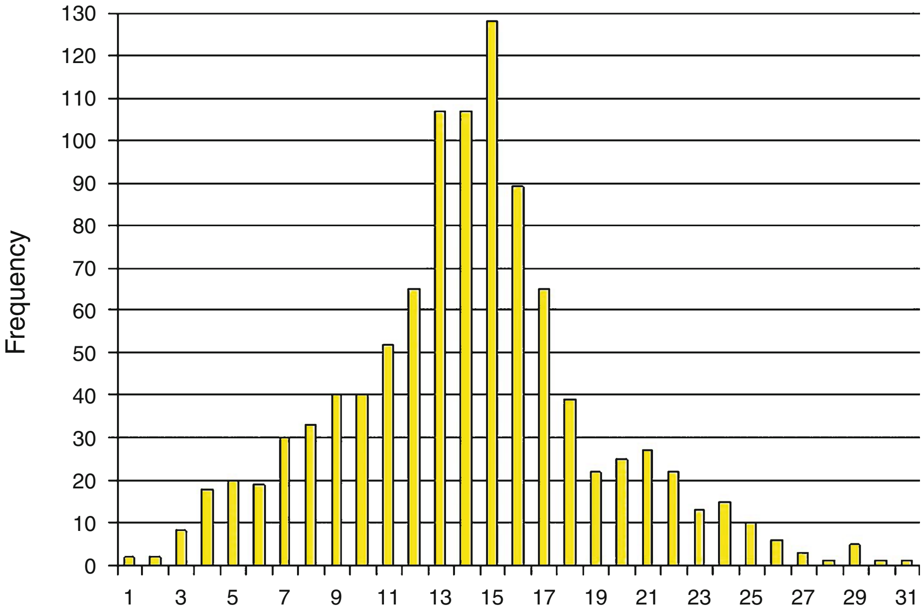

Fig. 16. Graph showing age at primary surgery with a median age of 14 years. Age ranged from 1 to 31 years at our institution

tions. Of the 86 patients undergoing redo procedures, 39 had a failed Ravitch, 44 had a failed Nuss, 1 had both a failed Ravitch and Nuss procedure and 2 had failed Leonard procedure. Of the 1,015 patients having primary repair, 690 patients have had their bar removed. There have been numerous important modifications which have been made both to the surgical technique (e.g., routine use of thoracoscopy) and to the instruments since the origination of this procedure. This has served to facilitate insertion and stabilization of the substernal support bar. These have markedly reduced the risks and complications and have been well documented in recent publications [19].

The male to female ratio in patients undergoing repair was 4:1. The median age was 14 years, with a range of 1–31 years (Fig. 16). The median Haller CT index was 4.6 with a range of 2.4–32.4. Cardiac compression was noted on echocardiography and/or CT scan in 793/889 (89%) patients. Mitral valve prolapse was noted in 132 (15%) patients. Resting pulmonary function testing (PFT) was completed in 900 patients and demonstrated abnormalities in up to 45% of the patients.

6.3.7 Operative procedure, analgesia, and length of stay

A single bar was inserted in 730 (71.9%) patients. Two bars were inserted in 281 (27.7%) patients. Four (0.4%) patients received three bars. Blood loss in most patients was minimal (±10 cc). The median length of stay (LOS) was 5 days with a range of 3–14 days.

6.3.8 Complications

6.3.8.1 Early complications (Fig. 17)

There were no deaths ($n = 0$) nor were there any cardiac perforations ($n = 0$) during the 1,101 primary and secondary repairs performed at our institution. It was common to have pneumothorax after repair but usually it resolved spontaneously. Pneumothorax requiring chest tube drainage occurred in 36 (3.6%) of the primary repairs and required only percutaneous aspiration in 3 (0.3%) primary repairs. In the redo population, 29 (34%) patients required a chest tube placement for evacuation of the pneumothorax with 2 (3%) resolving with aspiration only. Hemothorax requiring drainage but no transfusion occurred after four (0.4%) primary repairs. Three (0.3%) pleural effusions required treatment by either chest tube or aspiration.

In the population of primary repair patients, pericarditis requiring treatment with indomethacin or prednisone occurred following five (0.5%) repairs, with one requiring pericardiocentesis. Pneumonia occurred after 6 (0.6%) repairs, and medication reactions have occurred following 36 (3.6%) repairs. One

• Pneumothorax w/spont. resolution	60.4% ($n = 613$)
• Pneumothorax w/chest tube	3.6% ($n = 36$)
• Horner's syndrome	17.7% ($n = 179$)
• Drug reaction	3.6% ($n = 36$)
• Suture site infection	1.0% ($n = 10$)
• Pneumonia	0.6% ($n = 6$)
• Pericarditis	0.5% ($n = 5$)
• Hemothorax	0.4% ($n = 4$)
• Pleural effusion (requiring drainage)	0.3% ($n = 3$)
• Temporary paralysis	0.1% ($n = 1$)
• Death	0%
• Cardiac perforation	0%

Fig. 17. Early postoperative complications of primary surgical patients

hundred and seventy-nine (17.7%) patients had transient Horner's Syndrome at varying times during the thoracic epidural administration.

6.3.8.2 Late complications (Fig. 18)

Fifty-eight (5.7%) patients have experienced bar displacement, and 43 (4.2%) displacements warranted repositioning. After the introduction of stabilizers, the incidence of bar displacement dropped from 8.9% to 2.3%. There has been only one bar displacement (0.1%) since we combined placing a stabilizer on the left and PDS sutures around the bar and underlying rib on the right. Bar infection occurred in 11 patients (1.1%) requiring early bar removal in 2 (0.2%) patients. Twenty-nine (2.9%) patients had allergies to the bars. These presented in a variety of ways. Several patients gave a history of metal allergy and therefore received a titanium bar, stabilizer and wire suture. The others were diagnosed postoperatively and were treated with antibiotics or steroids. Three of these patients did not respond to medical treatment and required early bar

• Bar displacements	58/1015 (5.7%)
• Requiring revision	43/1015 (4.2%)
• Overcorrection	32/1015 (3.2%)
• Bar allergy	29/1015 (2.9%)
• Wound infection	11/1015 (1.1%)
• Recurrence	8/1015 (0.8%)
• Hemothorax (post-traumatic)	2/1015 (0.2%)
• Skin erosion	1/1015 (0.1%)
• Accidental death	1/1015 (0.1%)
(accidental death 3.5 years post op)	

Fig. 18. Late postoperative complications of primary surgical patients

removal, two of these received titanium bars. The symptoms resolved after removal of the steel bar. Thirty-two (2.9%) developed a moderate overcorrection of their deformity and four (0.4%) developed a true carinatum deformity.

6.3.8.3 Long-term follow-up

Patients are followed at 6 months postoperatively, and then yearly. Long-term assessments classify the post-operative results as excellent, good, fair, or failed.

A result is considered to be excellent if the patient experiences total repair of the pectus excavatum as well as resolution of any associated symptoms. A good result is distinguished by a markedly improved but not totally normal chest wall appearance and resolution of any associated symptoms. A fair result indicates a mild residual pectus excavatum without complete resolution of associated symptoms. And a failed repair indicates a recurrence of the pectus excavatum and associated symptoms and/or the need for another repair after bar removal.

The initial cosmetic and functional results at the time of repair were excellent in 938 (92.4%), patients, good in 74 (7.3%) patients, fair in 1 (0.1%), and failed in 2 (0.2%). Of these patients, 690 (68%) have had their bars out for more than 1 year. The results of the patients who had their bars removed one or more years ago are excellent in 587 (85.4%); good in 75 (10.9%); fair in 9 (1.3%), poor in 7 (1.0%) and failed in 9 (1.3%). There were three patients who did not return (Fig. 19). Patients who did not comply with the exercise program and had their bar removed before puberty had a higher recurrence rate. The length of time the bar was left in situ had a direct effect on the long-term outcome. The longer the bar stayed in, the better the results. The age of the patient also affected the long-term outcome with the best results occurring in the 7–12 and 13–18 year age groups. The long-term results of patients who have had their bar removed before December 31, 2006 show that

• Total number of primary patients	1015
• Total number w/bar removed	690 (64.2%)
– Excellent result	587 (85.4%)
– Good result	75 (10.9%)
– Fair result	9 (1.3%)
– Poor result	7 (1.0%)
– Failed	9 (1.3%)
– No return (bar removed else w)	3 (0.4%)

Fig. 19. Overall results after removal of pectus bar

Number w/bar removed prior to 12/31/06	587
– Excellent result	493 (84.0%)
– Good result	69 (11.8%)
– Fair result	9 (1.5%)
– Poor result	7 (1.2%)
– Failed	9 (1.5%)
– No return	2

Fig. 20. Long-term results in patients having their bar removed before December 31, 2006

we have 587 total patients with and excellent result in 493(84%) patients (Fig. 20).

6.3.9 Conclusion

The minimally invasive procedure provides good to excellent correction of pectus excavatum in over 90% of patients with no rib resection, no sternal osteotomy, minimal blood loss, and rapid return to normal activity. Studies have shown marked improvement in the patient's body image and have also shown slight improvement in pulmonary function [20]. The 1,101 patients managed at our institution have had excellent long-term results and low morbidity.

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6.4 Special considerations in adults with MIRPE and MOVARPE techniques

Anton H. Schwabegger

The deformity continues to worsen until skeletal growth has been completed in late adolescence and then changes little througout adult life.

Eric W. Fonkalsrud

According to but without knowledge of that fundamental citation of Fonkalsrud [10], many patients and their parents are not adequately informed about the opportunity to correct skeletal deformities of the anterior thoracic wall during growth and body maturation at an age, when surgical repair does not yet mean a major invasive and technically demanding intervention due to skeletal elasticity. Fonkalsrud in 2000 stated that only 15% of all patients affected by the deformity undergo surgery. With the advent of the MIRPE, this percentage initially increased, but on the other hand, also a significant number of present adult patients out of the remnant 85% thus untreated patients now seek for feasible modern treatment.

In almost no case a deep funnel diminishes with increasing age or growth spurt, yet the opposite is the case, as costal cartilages elongate with body maturation thus lead to increasing folding up (keel chest) or down (funnel chest) of the adjacent sternum [12, 37]. While the main advantage of the MIRPE in children is reshaping the anterior thoracic wall without osteochondrotomies and hence allowing for normal growth of the thorax, this advantage of pliability no longer is present in the adult thorax. Herein the size of the thoracic cage is already determined by the rigidity of the skeletal structures and musculature, thus the matured skeletal structures will not simply tolerate any remodeling forces just by transmission of pressure.

The flexibility of the thorax and in particular of the chondrosternal zone at the anterior thoracic wall in children represents the fundamentals for the success of the MIRPE method (Fig. 1a–f). On the basis of this flexibility, the method was developed for the correction of the juvenile pectus excavatum deformity by the pediatric surgeon Donald Nuss (Chapter 6.3). Remarkably he published the first series in 1998 [26], not prior to a very long observation period of up to 10 years, presenting successful and secured long-term results. The incontestable successes of this method now also made many adults, who were concerned by their unpleasant aesthetic burden, attentive to pull this minimally invasive method into consideration as a new option. They apparently preferred this concept with minor scars in contrast to open surgery techniques [6–8, 24, 33, 40] or alternatives such as autologous microvascular tissue remodeling [14, 16, 21, 25, 32, 38], well known so far. Osteochondrotomies in adults will not lead to growth disturbances any more, but may result in other problems like formation of scars in a non concealable area of the body surface, particularly in women and furthermore reduced flexibility of the thoracic wall.

Because of the success with children and youth, multiply published through many scientific papers [12, 26, 31] but moreover in public media, the desires and even demand [19] for the application have markedly increased also in the adult males and females equally [2, 3, 11, 20, 30, 35, 42].

Utilizing the pure MIRPE in adulthood however arise other and substantially more particular problems contrarily to an early age. The standardized fabrication of the pectus bar in uniform material strength and width on the one hand allows the use of instruments of different manufacturers. It furthermore turned out that this uniformity of instruments is advantageous especially for the explantation procedure of the pectus bar after years. Adult patients nowadays hardly any more remain stationary and due to increasing requirements of migration across nations they undergo treatment wherever it appears suitable. Therefore, more frequently pectus excavatum patients present themselves, primarily treated elsewhere or even abroad and claim to have the bars removed at the end of the implantation period after several years. Fortunately through the uniformity of the pectus bars and the instruments though fabricated by several manufacturers, this migration of patients until now yields no problem. For exactly these circumstances

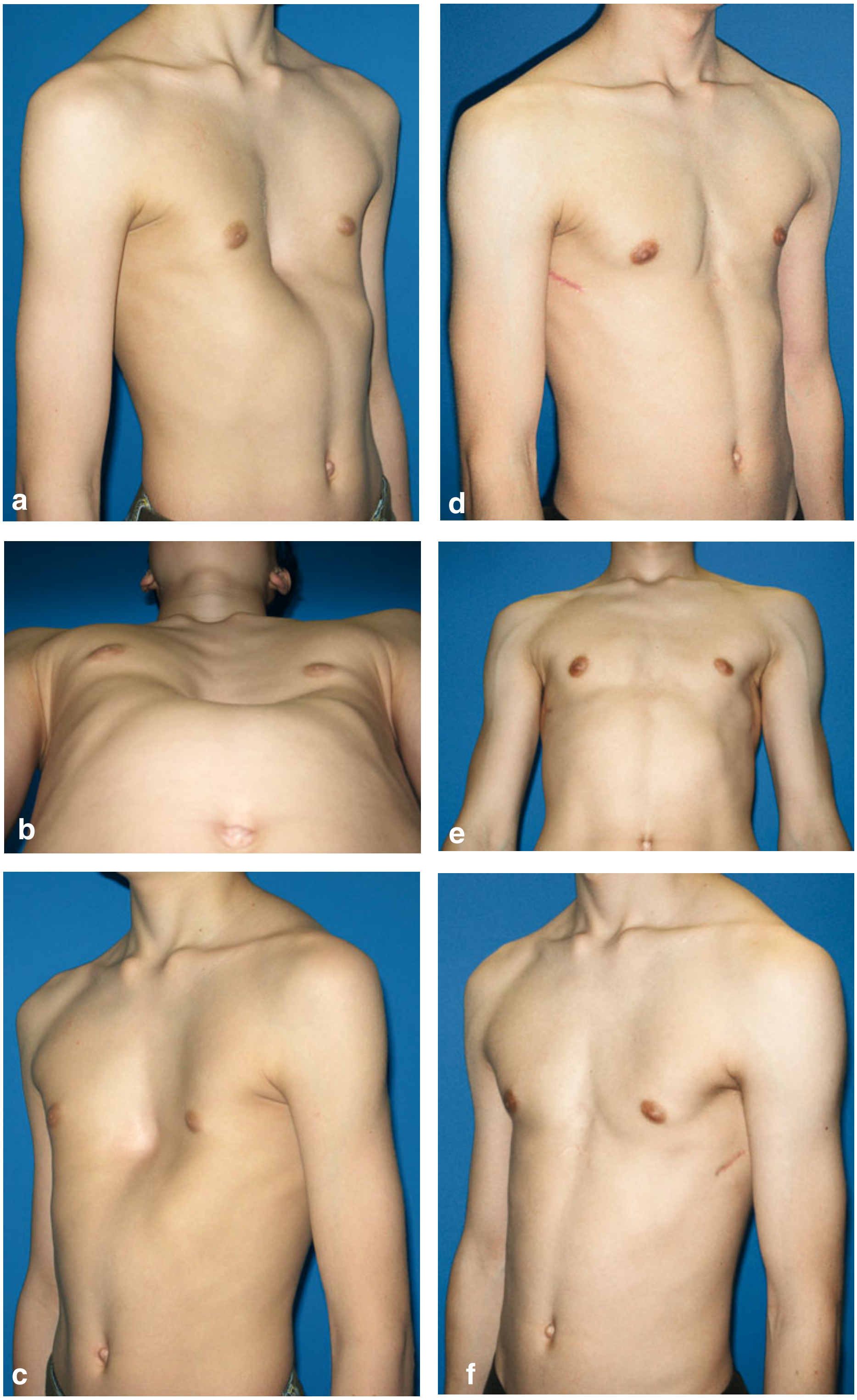


Fig. 1(a–f). 13-Year-old male with moderately asymmetric funnel chest deformity (**a–c**). Situation at an age of 17 years and 1 year after explantation of the pectus bar (**d–f**)

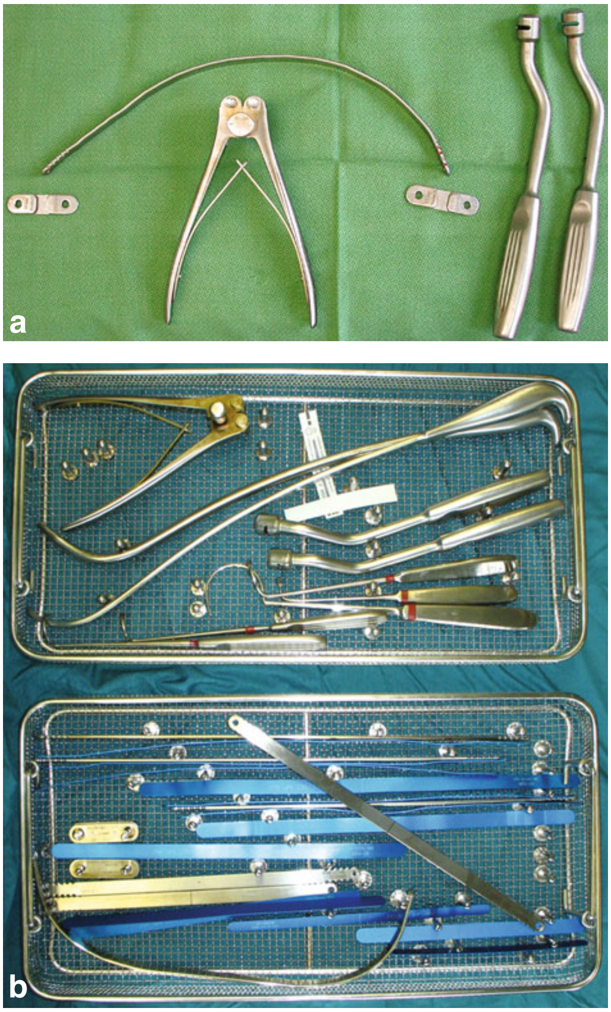


Fig. 2a and b. Standardized instruments for pectus bar implantation and explantation available from different manufacturers

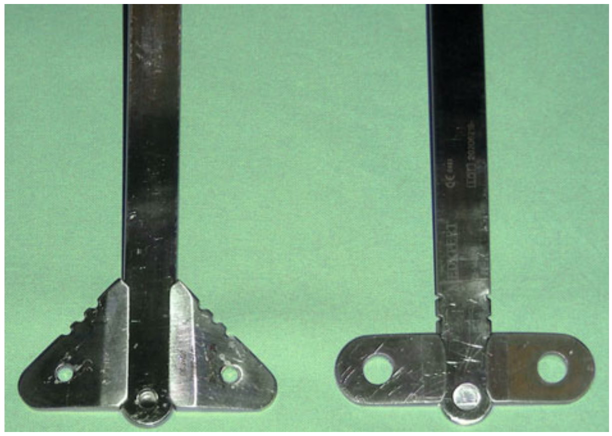


Fig. 3. Different shapes of lateral stabilizers, however fitting to the standardized pectus bars

of globalization it is to be hoped that the standard measures of the pectus bars and the relevant instruments (Fig. 2a and b) are furthermore retained uniformly within a consensus of different manufacturers and the applying physicians as well.

By this standardization on the other hand, boundaries within the use of these tools and materials are reached soon in the application in adults, because the material strength (thickness) of the pectus bar had been conceived and made just for the application in children. The massive pressure to the sternum in an exemplarily athletic adult with a deep funnel deformity therefore becomes too strong [9, 41]. Such differences in adults versus children have been measured and it turned out that the different load in athletic adults may reach 30-fold the load compared to a child [9]. It is therefore evident that even frequently suggested implantation of two pectus bars may rather not overcome the withstanding pressure from the rigidity of an adult anterior chest wall and other measurements have to be considered to alleviate that strain. In other words, the immense forces will be a subject of creating much more problems during surgery and postoperatively, in terms of difficulties of and during the surgical maneuver itself with increased rates of ensuing complications, whether immediately or later [8, 20, 22, 23]. The high pressure load focused at the back surface of the sternum on the one hand leads to partial unbending of the pectus bar in situ so that a major part of the supporting effect goes lost, if not primarily overcorrected. On the other hand, this unbending leads to sprawling of the bar wings laterally which cannot be prevented even with fixation techniques that use special stabilizers of different designs (Fig. 3). These laterally potentially projecting bar wings are then unpleasantly palpable (Fig. 19 in Chapter 12.1) and even well visible when attached with stabilizers. Moreover the high pressure on the vertex of the pectus bar leads to a pressure redistribution on the funnel margins and therewith onto the ribs, upon which the pectus bar rests. With such resulting high pressure load also at the back surface of the sternum and on the ribs, it becomes intelligible that pains are increased and therefore complained much more by adults than by children. Therefore, the MIRPE method was modified to use even two pectus bars for the purpose of load distribution [5, 23]. This modification has proved to be advantageous in certain indications such as a wide and flat funnel deformity, asymmetry [29], steep and deep funnel depressions; however it tends to create more complications yet [9]. Arrosion of the bony integrity of sternum or ribs with pathological fractures (Chapter 12.1), a prolonged pain symptom complex, and other consequences of such an expanded surgical manipulation are possible sequelae. These include increased risk of intrathoracal organ injury, increased inclination to pleural effusion, hematoma, or infection through an extended thus tissue damaging surgical access.

With all these and possibly other problems a surgeon is confronted, who tries to adapt this technique, which was primarily developed for children, unreflected to adults. The original MIRPE technique is extremely suitable for children and herewith already established for many years now [31]. If utilized in grown-up patients with much more matured and strengthened body, it will evidently bear more risks for complications or an undesired outcome [20]. Though being unprepared to and unaware about a totally altered anatomical and functional situation with different strengths of withstanding forces to remodeling attempts, such surgery might become a cumbersome endeavor. The overall biomechanics and possible compensation mechanisms basically differ from those in children [9, 31]. It is therefore unavoidable that in the initiation phase to apply the MIRPE also in adults, several shortcomings are made, furthermore particularly solely with adults no evidence-based reports of experiences with larger series and consistently selected patients yet exist.

However and on the other hand nowadays it is no longer possible to design prospective comparative studies between conventional techniques and the MIRPE technique [24]. Patients on the basis of their knowledge and the resulting prejudice, which is propagated by many media, can no longer be randomized and distributed into groups. The gain of experience often relies only on results obtained by individual case series published with short-term results only and anecdotical transmission of tips and tricks, however also experienced pitfalls. Several institutions perform corrective surgery but each with a variety of techniques, different surgeons, and overall within a collective of patients with inconsistent age, body shape, and extent of deformity. Therefore, no sufficient level of scientific evidence based on randomized controlled trials is available for the selection of individual proper treatment, especially for the treatment in adults. With increasing pressure through those who usually know well about case stories informed patients and probably increasing security in managing the peculiarities of the treatment in adults using the MIRPE technique, more reports containing larger series [15, 24] will hopefully become available. Equally long-term observations several to many years after the removal of the pectus bars are necessary to prove the benefit of the technique in adults too (Figs. 4a–g and 5a, b). The gain of experience in adult patients is still positioned in the status of a work-in-progress. On the basis of lacking and secured results with long-term observation at the current moment, one cannot yet state with certainty that the MIRPE technique applied in adults will provide equal success as that in children and adolescents, although several purely technical notes and reports with good results after short-term follow-up are already available, however already pointing out particular difficulties concerning technique and complication rates [20, 22, 23, 27, 28, 30, 35, 42].

Also the factor of yet ongoing growth before reaching adulthood still has an important influence on the success of the MIRPE. Its effects on the chest wall guide the remodeling process by a pectus bar during childhood and substantially better in adolescence. The conversion forces from cavity to convexity at the sternum and adjoining ribs by the pectus bar should be set at a time that transmission of forces is accompanied by skeletal growth. As such biodynamics through guided growth by the pectus bar over years lead to a stable formation of the anterior thoracic wall. Such a guided shaping evidently should lead to stagnation of any remodeling forces after maturation of body growth. Therefore even after removal of the pectus bar sufficient rigidity has developed and no further disfiguring growth of rib cartilages may occur. Just these biomechanical considerations now vote for putting the surgical correction of asymptomatic funnel chest deformities by MIRPE into puberty, on the one hand in order to utilize guided chest wall formation through finalizing maturation growth and on the other hand in order to obtain also a stable and therewith durable result.

Under these aspects, the use of MIRPE appears rather inopportune at the adult skeleton because no shaping growth guided by hardware takes place any more. The realization of conversion from a cavity to arching herewith is managed through the pressure of the pectus bar to the sternum. In analogy to osteodistraction, an osteo-chondro-distension of the anterior thoracic wall takes place. Such forces considerably necessary in tall or athletic adults can hardly be held by a single pectus bar. Occasionally a second pectus bar is sufficient, but at times even that modification will not suffice to withstand the biomechanical forces caused by rigidly disfigured anatomy. This problematic nature is the more stronger and steeper the funnel deformity is, the more athletic and older a patient is. The pure MIRPE yet can be the method of choice in a 35-year-old slim woman (Chapter 9), but can present a contraindication in a sportive 20-year-old fellow, on the basis of complications to be ex-

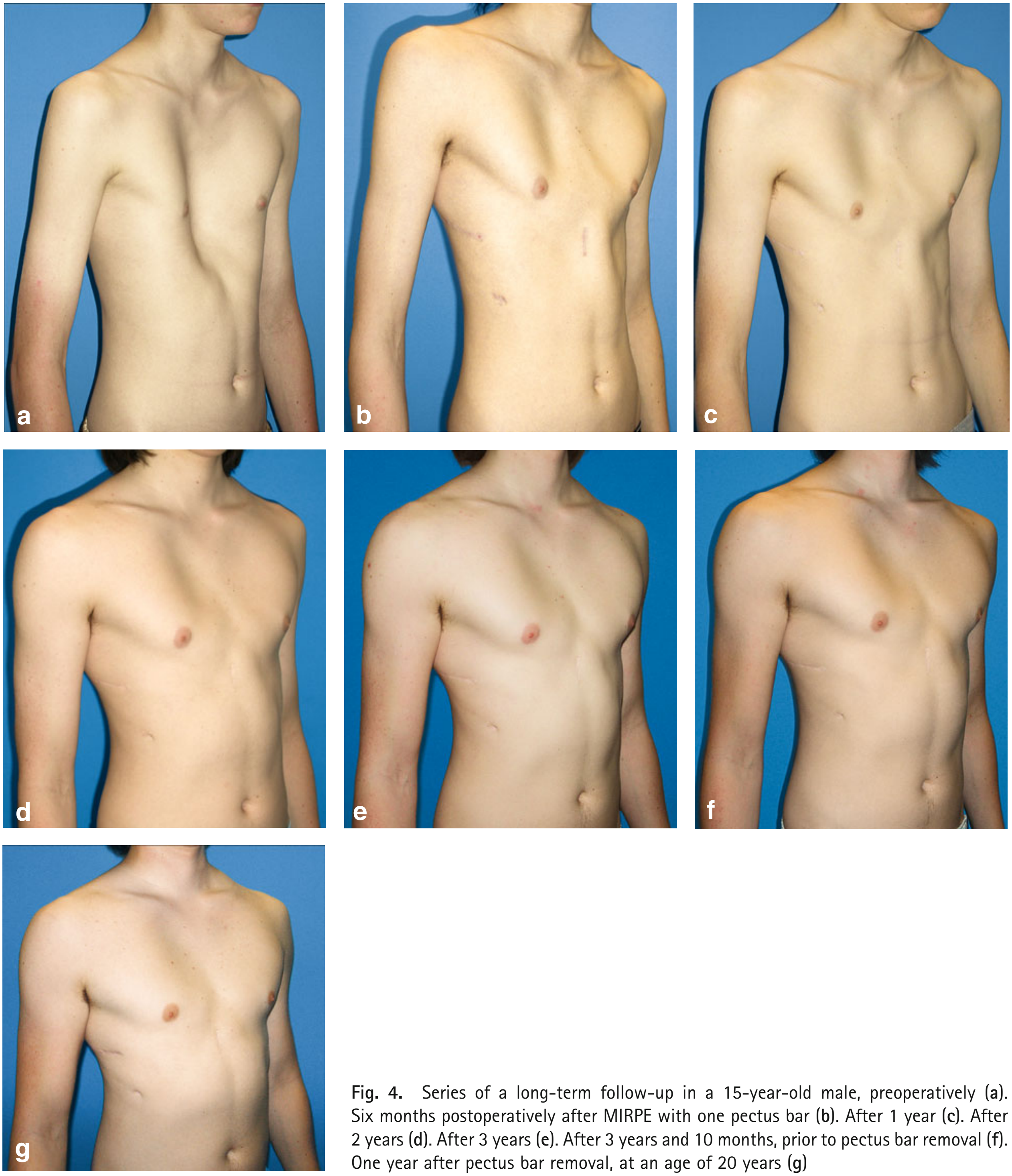


Fig. 4. Series of a long-term follow-up in a 15-year-old male, preoperatively (**a**). Six months postoperatively after MIRPE with one pectus bar (**b**). After 1 year (**c**). After 2 years (**d**). After 3 years (**e**). After 3 years and 10 months, prior to pectus bar removal (**f**). One year after pectus bar removal, at an age of 20 years (**g**)

pected due to biomechanical strains through muscular tightness and skeletal rigidity.

In such cases inappropriate for the MIRPE a modified combined access with osteochondrotomies performed via additional surgical access may be better suitable. The pectus bar implantation is supported through a complementary central access at the submammary or parasternal area to perform a horizontal sternotomy at a level more cranially, with or without parasternal chondrotomies or chondrectomies, according to the method

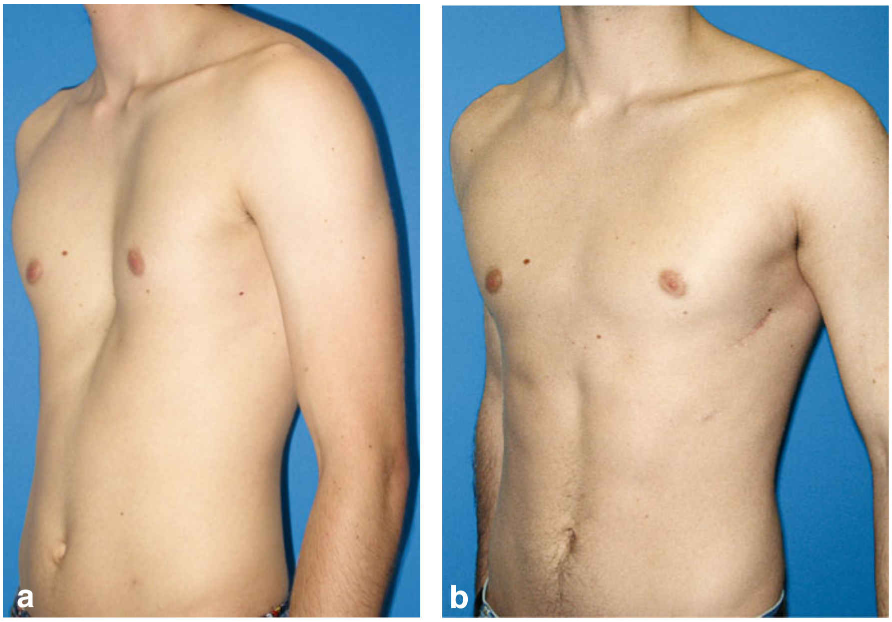


Fig. 5. 19-Year-old male with moderate funnel chest deformity, planned for MIRPE with one pectus bar (**a**). Situation 3 years after pectus bar explantation with very pleasing result, now 6 years after initial surgery (**b**)

of Ravitch (Chapter 6.1). Al-Assiri in 2009 noted in a comparative study, however in a collective of children, that the addition of sternocostal relaxing incisions to the standard Nuss procedure appears to facilitate retrosternal dissection and bar placement [1]. However, there were no changes in long-term function or cosmesis notable. The use of relaxing incisions also in children appears to be safe and may facilitate operative visualization of retrosternal structures. This concept of relaxing incisions but with additionally required chondrectomies in the rigid adult skeletal structures seems to expand the feasibility of minor invasive remodeling of the anterior chest wall to mature bodies in adults and athletic or tall patients within adolescence likewise.

6.4.1 Surgical technique with the modified hybrid access in adolescents and adults, the MOVARPE (Minor Open Videoendoscopically Assisted Repair of Pectus Excavatum) technique

The initial steps of surgery in the MOVARPE technique are the same as in the MIRPE technique (Chapter 6.3), but hybridized with additional incisions and remodeling interventions according to the combined technique published by Jensen in 1970 [17]. Bilateral incisions expose the serratus anterior muscles, their slips are bluntly separated with utmost care not to damage the lateral thoracic nerve. The undersurface of the serratus muscle interdigitating with the pectoralis major muscle is dissected free from the chest wall and a tunnel until the funnel rim medially as well as a pocket for the bar wings

Fig. 6. Intraoperative situs showing the surgical incision placed along the submammary crease for the purpose of sternotomy using an angled saw and a specially designed retractor ready to fix a cold light source for better survey underneath the elevated skin

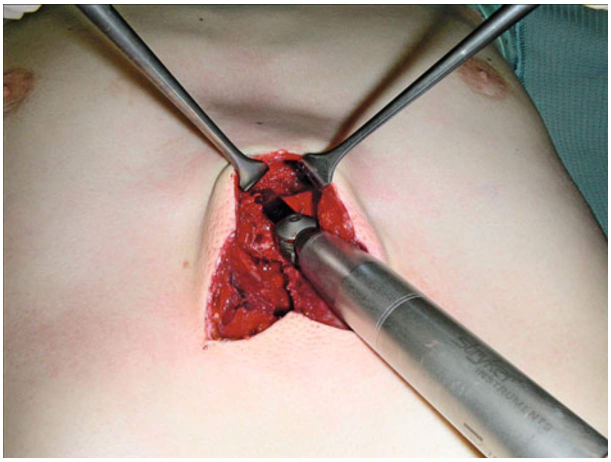


Fig. 7. Procedure of sternotomy in a male patient via a presternal incision

laterally is created. The incisions always follow the RSTL especially in the additional incisions at the anterior thorax. Here the incisions (Fig. 6) are placed near the planned relaxing chondrotomies and within a distance by which the sternotomy may be accomplished within a subcutaneous tunnel using an angled saw (Chapters 9 and 13.2). The pectoralis major muscle flaps may be raised or preferentially dissected in the muscle split technique to reach the individual ribs for chondroplasties [36]. Severely distorted rib cartilages are resected subperichondrially, whereas minor deformations are corrected by wedge resections or incisions just for weakening the continuity or relaxing the distorted arches of the ribs. The sternotomy (Fig. 7) is also performed as a wedge resection to create a small gap that enables elevation of the mobilized central thoracic wall unit. In symmetrically developed funnels, the osteotomy does not include the posterior table of the sternum in order to prevent dislocation by mobilization. Solely in the cases with malrotation of the sternum, the osteotomy has to be performed as a complete transsection to allow derotation of the sternum into a frontal plane. Either using a bone hook or using the finger after blunt separation of the pleura and pericardium from its back side, the sternum is elevated [17], causing intentional green-stick fracture of the posterior table of the sternum, if not totally transsected otherwise for the purpose of sternum derotation. Now the endoscopic ports are placed at the anterio-lateral chest wall about two or three intercostal spaces above the lowermost ribs and rib arch (Fig. 8). The tunnelizer under videoendoscopic view and deflation of the right lung (double lumen intubation,

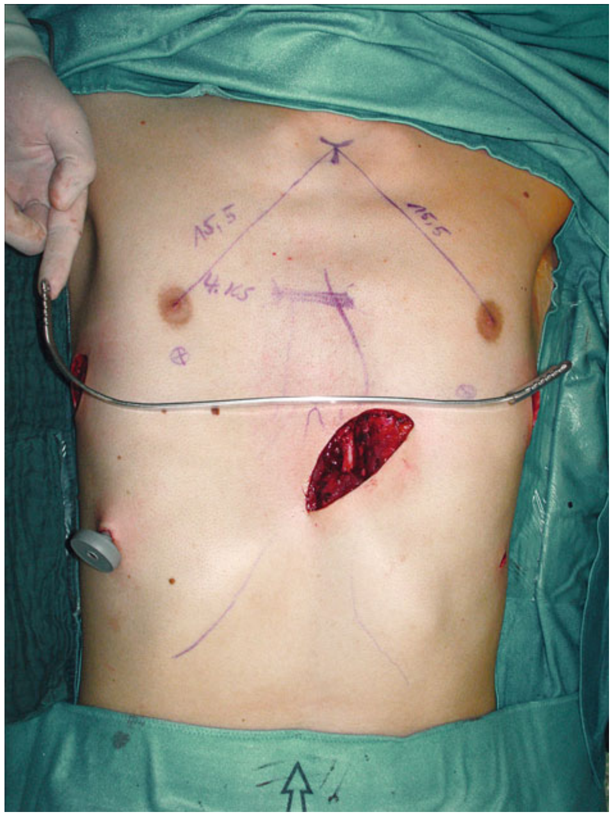


Fig. 8. Intraoperative view with already performed relaxing sternochondroplasties via the left parasternal incision. The shaped pectus bar placed into the desired direction above the skin. The endoscopic ports placed at the lower rib arches, ready for insertion of the endoscope

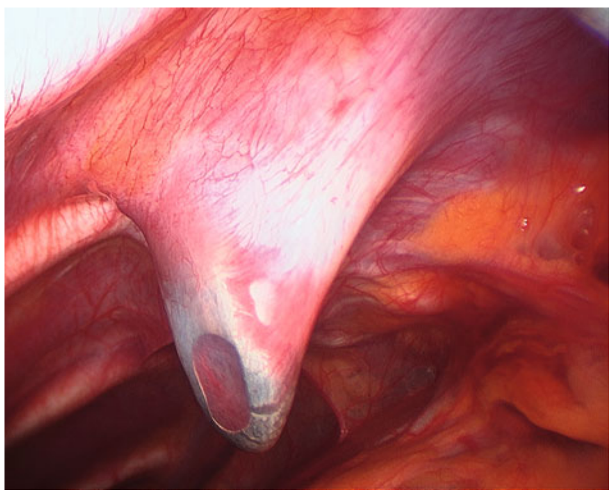


Fig. 9. Endoscopic view into the right thoracic cavity with the tip of the round tunnelizer entering the thoracic cavity prior to perforation of the parietal pleura

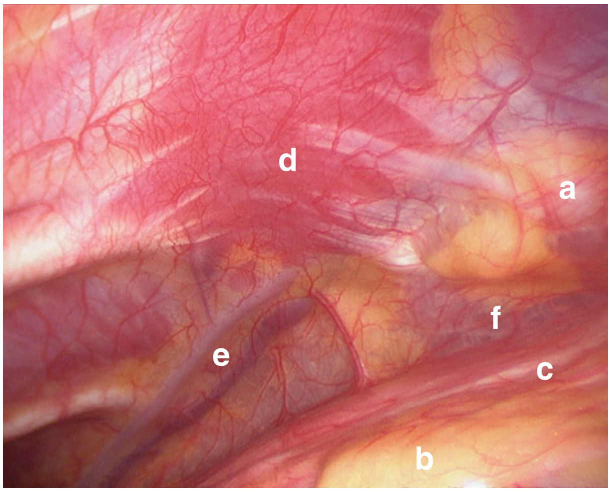
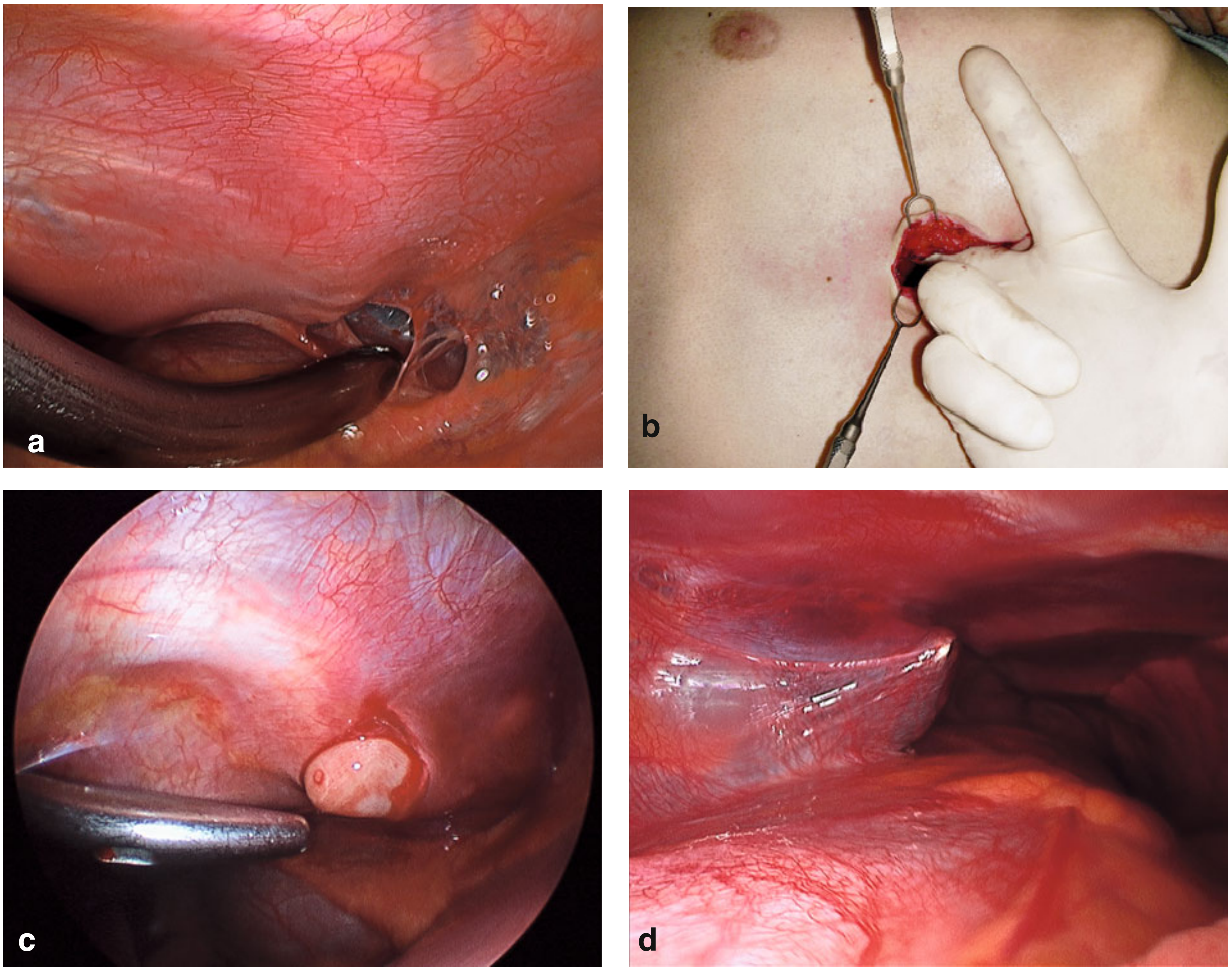


Fig. 10. Endoscopic view with posterior aspect of the right parasternal chest wall. (*a*) Sternum depression, (*b*) pericardium, (*c*) phrenic nerve, (*d*) transversus thoracic muscle, (*e*) internal thoracic (mammary) vessels, (*f*) point of retrosternal passage of tunnelizer and pectus bar

Chapter 5.3) now perforates the intercostal muscles and pleura to reach the thoracic cavity (Fig. 9). With simultaneous elevation of the mobilized anterior chest wall the tunnelizer is gently pushed versus the midline with particular caution to the heart, the phrenic nerve, and the internal thoracic vessels (Fig. 10). The chest wall elevation now alleviates the passage of the instrument (Fig. 11a) behind the sternum and in front of the pericardium [1, 5]. During that maneuver, the introduced finger simultaneously to visual control may guide the tip of the instrument between the mediastinal organs and the sternum (Fig. 11b and c). Under alternate insufflation and deflation of the lungs, the procedure is proceeded at the left throacic cavity again under visualization by the videoendoscope (Fig. 11d and e); the thoracic wall is perforated within the rim of the funnel likewise to the contralateral side. Due to the rigidity of the thoracic structures rupture of the intercostal muscles beyond the funnel rim caused by undue shoving and leverage forces of the tunnelizer means a probable technical failure. Furthermore such surgical violence will disrupt intercostal muscles from the ribs, either causing large holes in the pleura or causing immediate dislodgement of the pectus bar to dorsal, along the course of the ribs. Concomitant complications such as pneumothorax, wound infection, and late sequels like bar dislocation, and reduced respiration capacity by scarring or migration of the pectus bar into the thoracic cavity may occur (Fig. 17 in Chapter 12.1). That is why a rounded tunnelizer without edges is preferably used to avoid shearing-off effects (Chapter 13.2). The shaped pectus bar intraoperatively is fixed to the tunnelizer with a strong suture (e.g. Mersilene® strap), lubricated with gel, and then implanted in a reverse way. This maneuver is again performed under alternate ventilation, elevation of the anterior chest wall, and permanent visualization by the endoscope (Fig. 12a–f). After implantation, the pectus bar is tilted with the flipper instruments along its own axis to its definite position, with the bar wings placed into submuscular pockets at the lateral thoracic wall. Final inspection of the thoracic cavity by endoscope is done prior to wound closure (Fig. 12f). The bar wings are fixed with several circumcostal PDS-sutures (Fig. 13a–e) in order to avoid displacement [13, 39]. By the paramedian access, a third fixation parasternally but lateral to the internal thoracic vessels is feasible and recommended to prevent dislocation (Fig. 13f). During suturing the deep wound layers, the anesthesist is requested to raise the PEEP to extrude remaining air from the pleural cavity via the endoscopic ports until final skin closure. Commonly no chest tube then is necessary, although in most of the cases [4] a sickle-shaped small but asymptomatic pneumothorax is recognizable, which usually dissolves within a few days. Postoperative X-rays are recommended during the stay to exclude pneumothorax, hemorrage, or pleural effusion (Fig. 14a and b).

In summary, the main advantage in the application of a hybrid technique consists of the advantages of the rapid and elegantly performable MIRPE conjoined with parts of the conventional Ravitch procedure to allow a remodeling procedure without tension and stable support of the elevated anterior chest wall. Besides that, postoperative pain episodes are diminished by the fact that lever forces of the pectus bar versus the sternum are reduced by such additional relaxing and mobilization maneuver. On the other hand, the time needed for pectus bar support until healing of sternotomy and chondroplasties is notably reduced down to 1 year versus the period of up to three or more years in the chest wall distension technique of MIRPE. The surgical access to the sternum with relaxing osteotomy and parasternal chondroplasties allows stable scarring already after several weeks, supported by a pectus bar for 1 year then to enable further rigid healing. Such a formation of callus in the area of the sternotomy by secondary bone healing after few months sufficiently



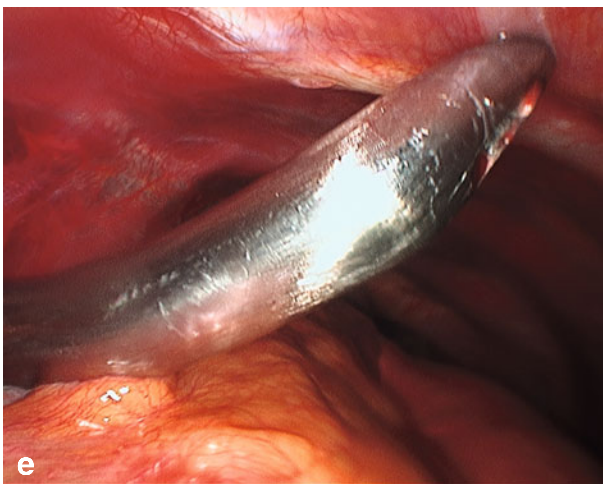


Fig. 11. Tip of the tunnelizer passes the pericardium just behind the sunken part of the sternum and gently dissects free the pleura from its back side (**a**). Simultaneous elevation of the relaxed sternum is achieved by the surgeon's finger via the presternal access, introduced behind the lower part of the sternum (**b**). Digital and endoscopic guidance of the instrument during sternum elevation and further passage of the tunnelizer (**c**). Passage of the tunnelizer tip to the left thoracic cavity, the endoscope now also changed to the left (**d**). The instrument is ready to perforate the left chest wall at the funnel rim (**e**)

allows permanent stability then (Figs. 15a–b, 16a, b and 17a–f).

The extension of the MIRPE to a MOVARPE technique in adults on the other hand increases the surgical expenses and furthermore the operation time by approximately up to an hour. An additional disadvantage is the need for one or two separate skin incisions, creating further scars. However,

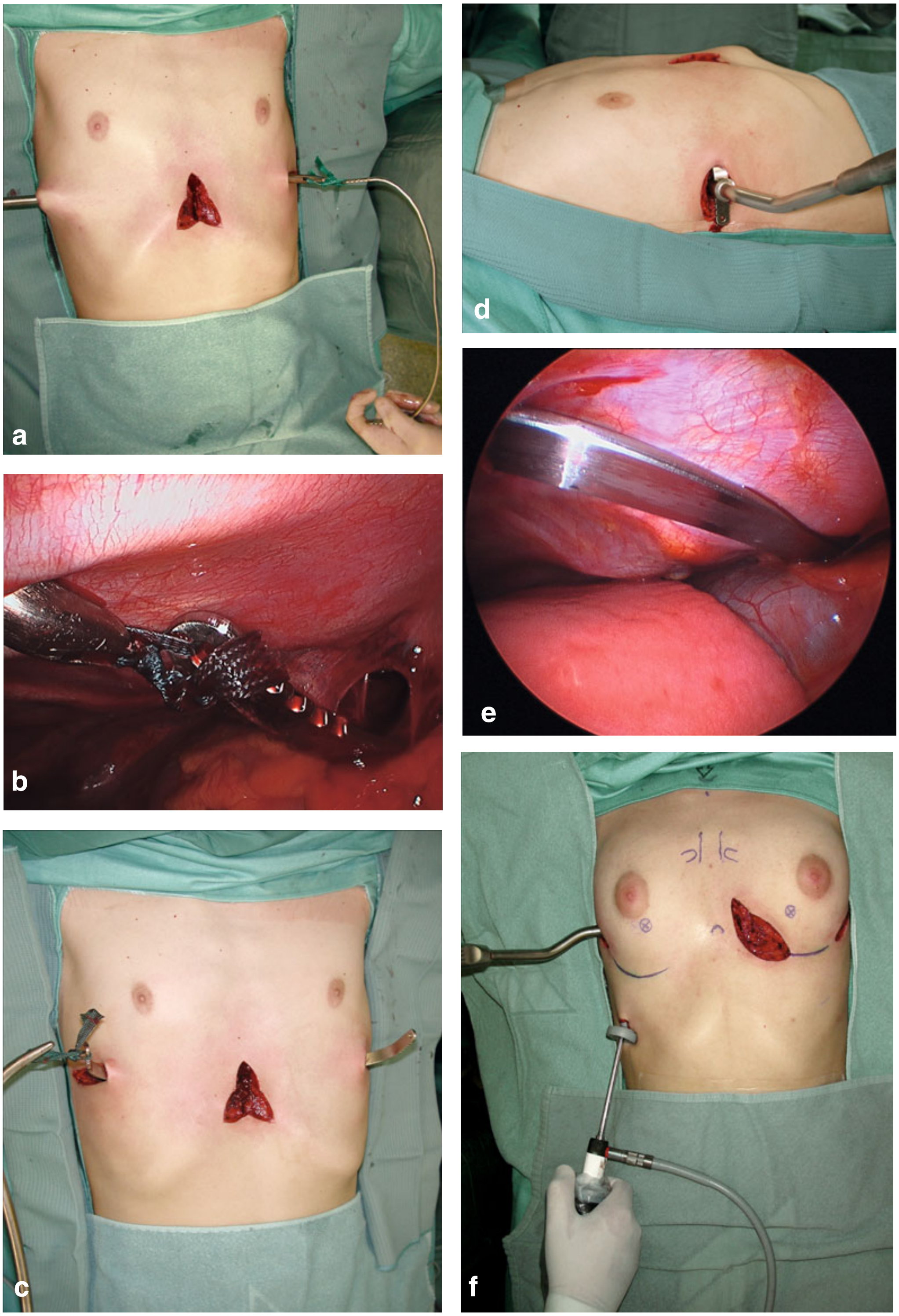


Fig. 12. Tunnelizer in place, the shaped pectus bar is fixed to the tunnelizer with a strong band (a). Endoscopic view into the right thoracic cavity, the pectus bar hitched to the tunnelizer already passed the left thoracic cavity, the retrosternal area, and pericardium, prior to emerging through the right chest wall (b). Passed pectus bar, still being tilted up prior to twisting it along a transversal axis (c). Utilizing the flipper instruments to twist the pectus bar at 180° and to place it into its desired final position (d). The pectus bar seen from the right thoracic cavity with apparent lever pressure against the posterior surface of the sternum (e). Inspection of both thoracic cavities after pectus bar tilting into its final position using the endoscope to exclude any unintentional trauma or complications (f)

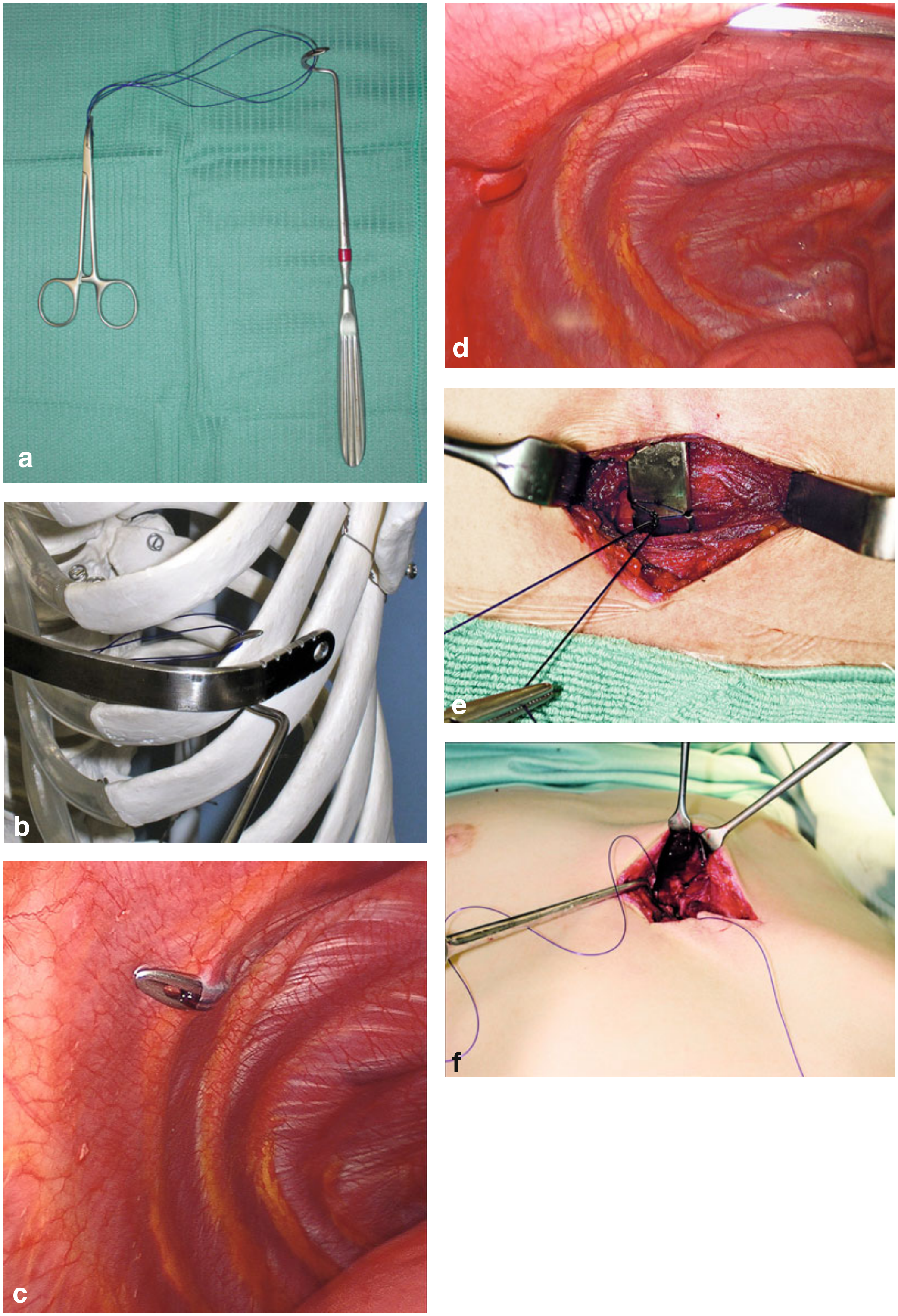


Fig. 13. Deschamps needle armed with PDS suture (**a**). Schematic depiction how circumcostal suture is placed with Deschamps needle and fixes Pectus bar wing to the rib to avoid tilting (**b**). Endoscopic view at the right lateral thoracic wall with invading tip of Deschamps needle, closely passing around the rib (**c**). Final situation with fixed suture (left) and pectus bar (above) in place (**d**). View at lateral incision and wing of pectus bar with circumcostal suture (**e**). Parasternally placed (three-point) suture for additional security against dislocation of the pectus bar (**f**)

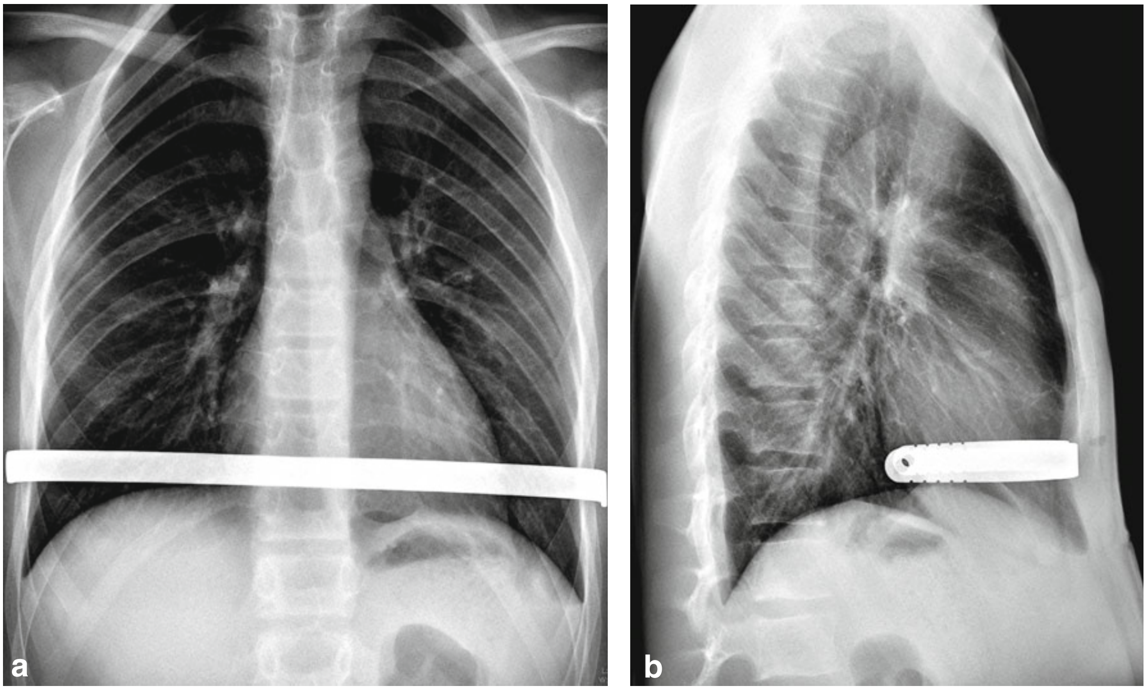


Fig. 14a and b. Postoperative X-rays with perfectly placed pectus bar, without hemothorax, hemorrage, or pleural effusion

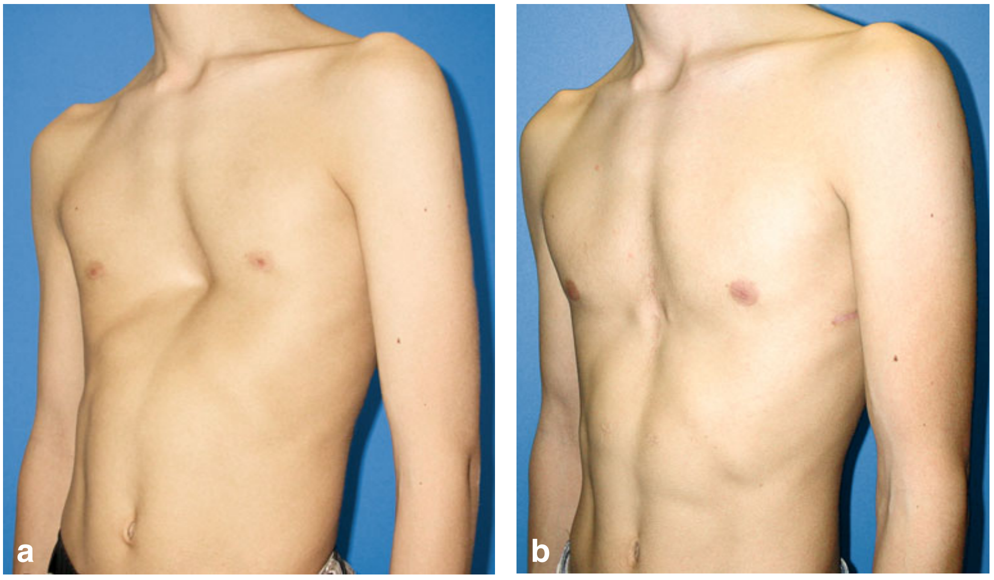


Fig. 15. 16-Year-old male with severe pectus excavatum deformity, psychic alteration, and moderate cardiopulmonal restriction (**a**). One year after explantation of the pectus bar, 2 years after initial surgery (**b**)

these scars may be placed distant from the midline and especially in females (Chapter 9) may be placed very well hidden along the inframammary crease (Fig. 12f).

6.4.2 Discussion

Results with the MIRPE in adults may be presented as good and excellent in so far available publications of

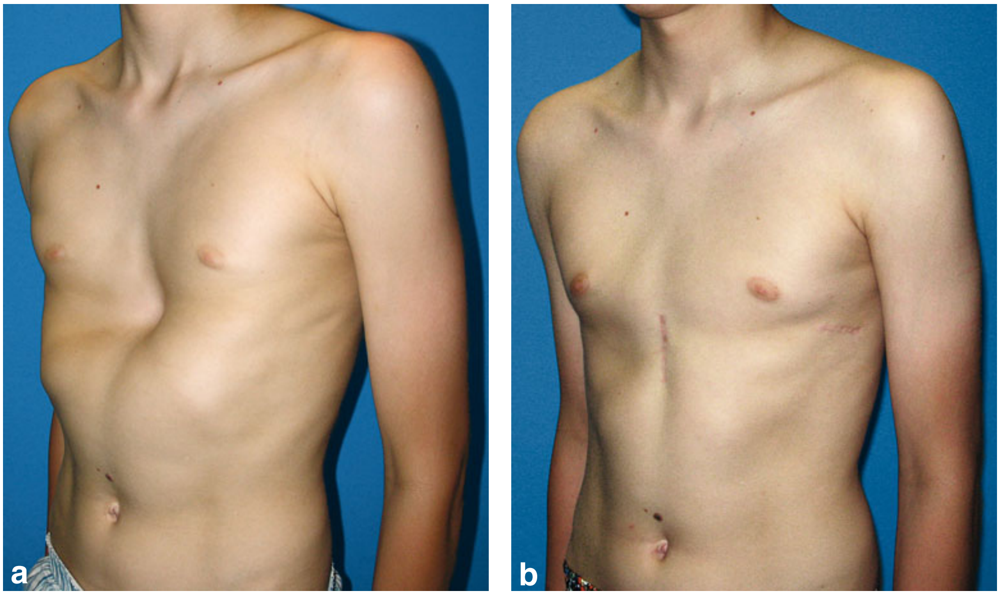


Fig. 16a. 15-Year-old male with severe funnel chest deformity but absence of function impairment. **b** Situation 1 year after pectus bar removal and 30 months after initial hybrid MOVARPE surgery

larger series in the Asian population [2, 5, 18, 22, 23]. However, it must be taken into consideration that Asians usually have a more asthenic physique than exemplarily Europeans. Weight over 90 kg and tallness above 190 cm in adolescents or adults within the caucasian race presenting with pectus excavatum are not rare incidents. The average physical characteristics of caucasian population with or without chest wall deformities usually lie far from those in the Asian population. That is why relevant publications with good outcome series in Asian adults [18] may not be simply transmitted without reflection to different biomechanical properties of the body and skeleton within different races. Leonhardt in a series with MIRPE in 48 European adults in 2005 pointed out that adults with severe or asymmetric deformities are at a greater risk of recurrence after a Nuss repair, thus 27% of these patients had to undergo reoperation with a Ravitch technique for finally content reshaping [20].

These findings in Europeans suggest that a hybrid technique using the MIRPE conjoined with partitions of the Ravitch procedure as the so-called MOVARPE technique will increase the general beneficial outcome when applied in adults. Until now, at our institution 21 selected patients (Fig. 18a–c) with a mean age of 24, 8 years underwent a hybrid repair of pectus excavatum and so far no patient required reoperation; minor relapse of sunken sternum (0.5–1 cm) was apparent in 4 patients only after pectus bar removal (Unpublished data, 2009).

6.4.3 Final comments

The MIRPE due to several advantages remains as the ideal therapeutic option in childhood and adolescence [4, 26], even in selected cases in adulthood. Nevertheless the more rigid and more severe the deformity appears beyond puberty, the hybrid technique MOVARPE seems to represent an alternative method with lesser pain periods, lesser pectus bar implantation period, and a lower rate of common complications, to be noted at first a tilting of the bar.

Above all in extensive pectus excavatum deformities at a higher age, also in existence of systemic diseases like Marfan syndrome or previous operations at the thorax, for surgeons of different disciplines it is very advisable to carry out indication setting and surgery as an interdisciplinary treatment jointly with thoracic surgeons experienced with that manner and potential intrathoracic complications.

Furthermore the indication for minimally invasive (MIRPE), minor open (MOVARPE), or conventional invasive surgery (Ravitch) at a body unit functioning for breathing must carefully outweigh the advantage of a psychological benefit.

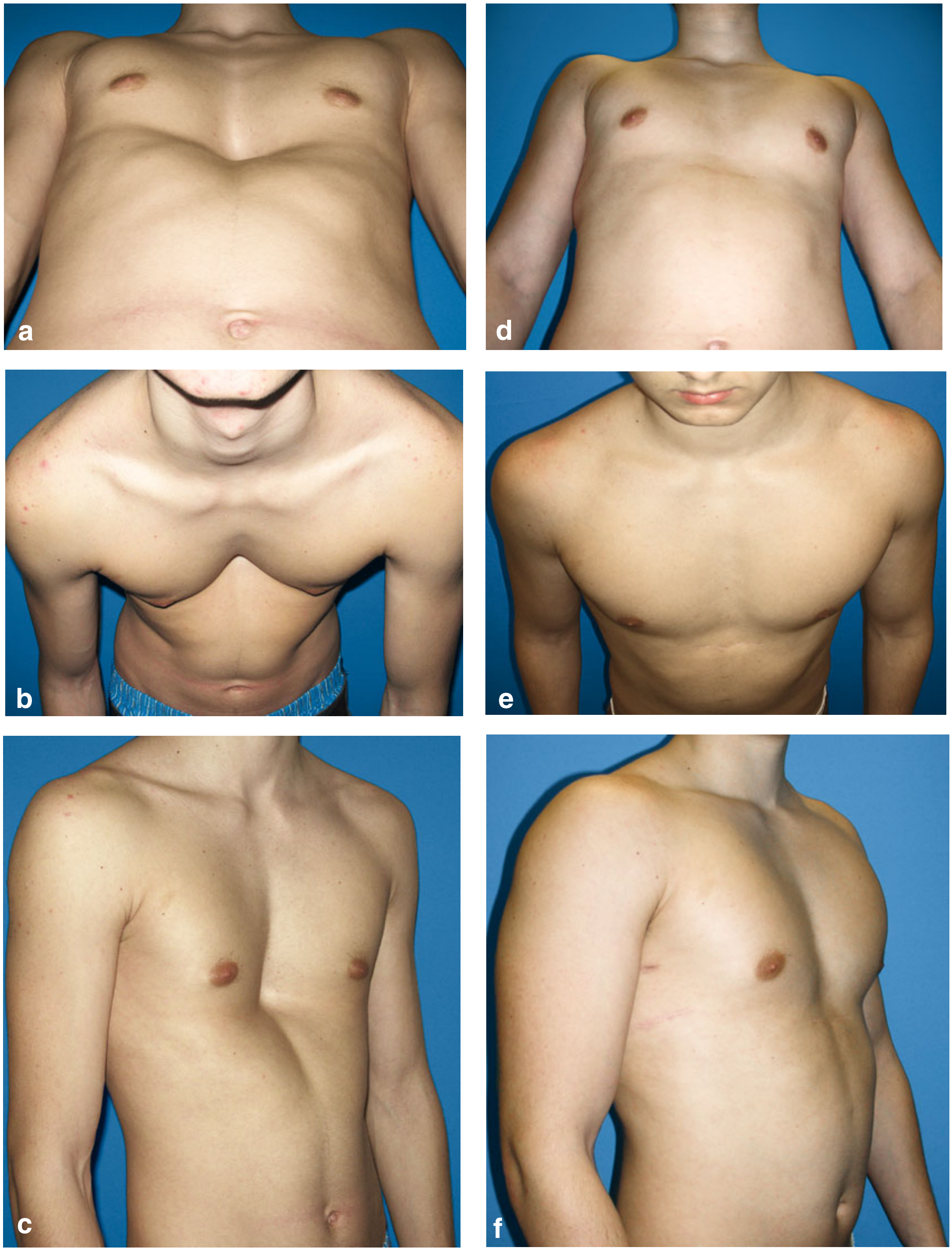


Fig. 17. 18-Year-old male with marked deformity and very rigid thorax (**a–c**). Situation 3 years after initial hybrid MOVARPE surgery. The patient refused removal of the pectus bar planned 1 year after implantation, it was finally removed after 3 years (**d–f**)

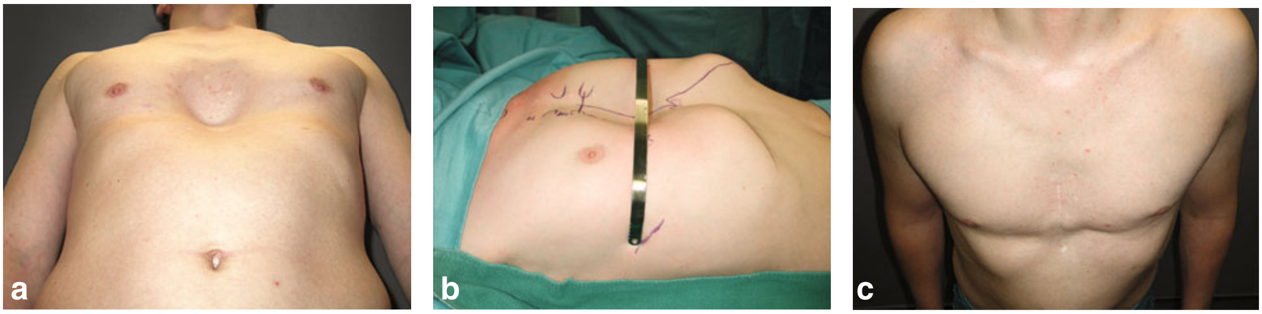


Fig. 18. 45-Year-old male with increasing cardiopulmonal impairment thus definite indication for anterior chest wall repair (**a**, **b**). Situation 1 year after pectus bar removal and 30 months after initial hybrid MOVARPE surgery (**c**). The cardiopulmonal function markedly increased

Surgeons taking care of adult patients also should be cognizant of all treatment modalities up-to-date and each intervention must be carefully selected and tailored to the individual patient.

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6.5 Custom-made silicone implants

Anton H. Schwabegger, Barbara Del Frari

6.5.1 Introduction

The method of simple implantation of a prefabricated, so-called custom-made implant made of silicone rubber has tradition since several decades already [1] especially for those cases, where a pure aesthetic improvement of the deformity in adults was aimed. In the seventies and eighties of the last century the development of an at room temperature vulcanizing [2–4] so-called RTV 382 silicone led to very pleasing aesthetic results with only one advantageously minor intervention and long-lasting results. That RTV silicone was implanted as a device vulcanized 24 h prior to surgery or as a viscous liquid component that vulcanized (polymerized) into a solid though elastic block in situ, within a dissected subcutaneous tissue pocket. By that and prior to the polymerization process in situ, the silicone temporarily contained viscous properties for some minutes, so that it was still feasible to perform simple manual contouring in situ with respect to the intended form and extension of the dissected subcutaneous pocket. The process of polymerization however, produced unpredictable reactions at the skin and subcutaneous tissue and thus was banned by the US government in the early eighties. Pretty often these implants nowadays, after two to three decades of unproblematic course, are removed due to calcifications within the tissue capsule and subsequent chronic seroma formation (Figs. 1–3). In the few cases of our own series of explantation however, no capsular contracture was found upon exploration during explantation. These implants still present themselves as pliable and soft like rubber gum (Fig. 4a, b). Due to the development

Fig. 1. Chronic seroma with reactive skin irritation lasting for over 4 months, 23 years after implantation of a RTV 382 at room temperature vulcanizing silicone for a pectus excavatum deformity. Bulging deformity and pain now indicated removal of the implant

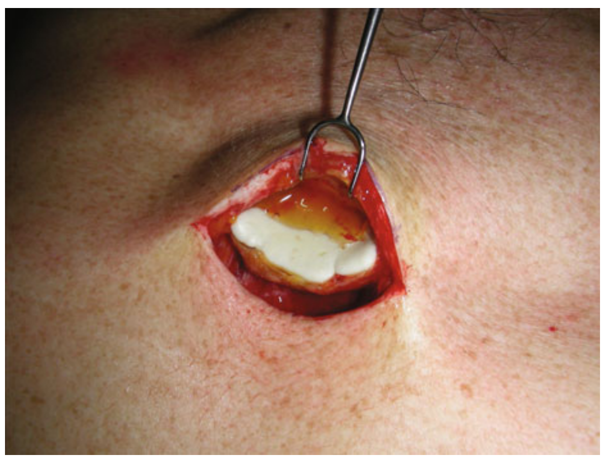

Fig. 2. The situation upon surgical exploration, the implant almost entirely floating in microbiologically diagnosed sterile seroma within the already opened fibrous capsule

Fig. 3. Tissue pocket after explantation of the whole implant depicting the capsule with calcification plaques (yellow) along most of the inner surface of the capsule

of prefabricated silicone implants, the utilization of that polymerizing silicone was abandoned soon thereafter. Nowadays, after several steps of technical improvements these implants are available and readily used [5–7], either solid and inflexible on the one hand, or very soft and pliable on the other hand, whether with smooth or textured surface (Fig. 5), with or without fixation tongues. Alternatively to such tongues, multiple holes may be cut into the implant using a surgical rongeur, enabling ingrowths of granulation and subsequently scar tissue, which provides with tight fitting and avoidance of dislocation (Fig. 6a, b).
Nowadays the technique of utilization of prefabricated or so-called custom-made silicone implants is very effective, minimally invasive and safe, with reproducible long lasting good to perfect aesthetic results and commonly very high patient satisfaction [8, 9]. It is a useful technique for the adult patients when growth has

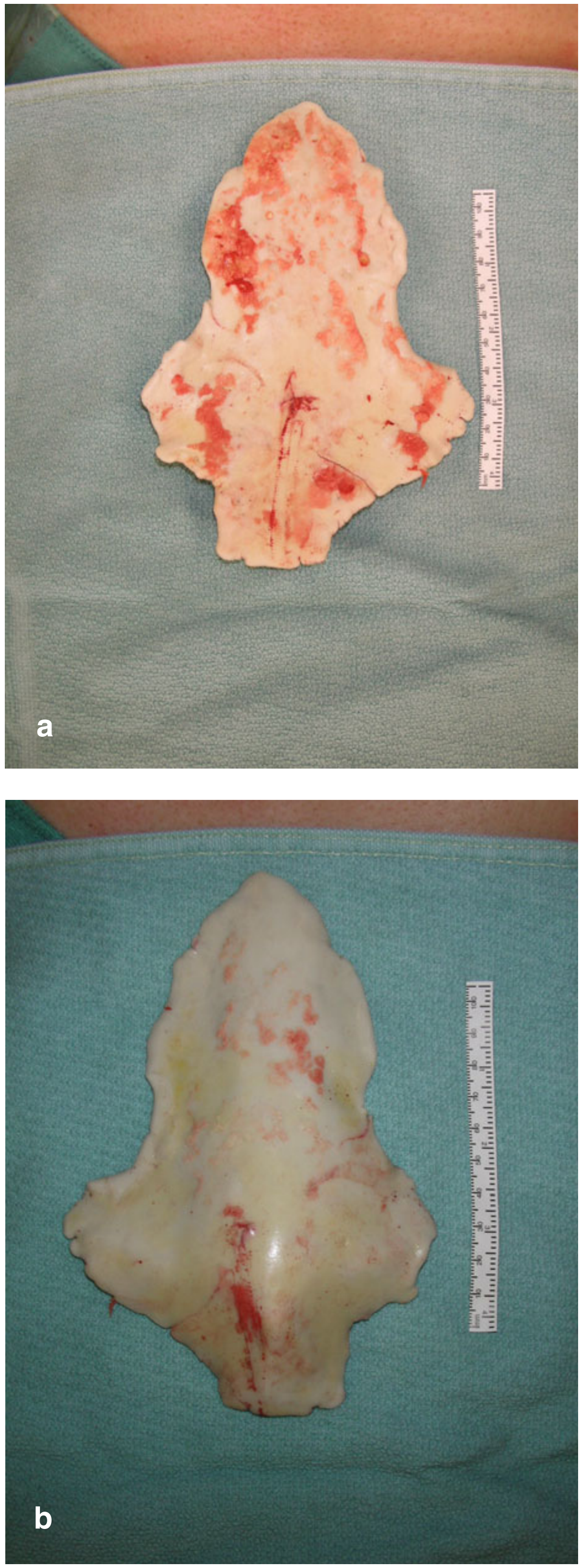

Fig. 4a. Anterior surface of the explanted RTV 382 silicone, same patient as in Figs. 1–3. **b** Posterior surface of the explanted RTV 382 silicone, same patient as in Figs. 1–3

Fig. 5. Custom-made modern silicone implants of various size, the upper right and the lower left with different kinds of textured superficial structure, the other implants show smooth surfaces

already finished (Figs. 7 and 8) and for patients with only a mild expression of the pectus excavatum deformity [7, 8, 14, 15] and particularly in patients with a Poland syndrome (Chapter 10.1). It is ideally suitable for asymptomatic patients with sternal depression but without cardiopulmonary problems [10].

Though, it must be taken into account, that after implantation of a moderate or unfavorable large-sized silicone block, a cramping anatomical deformity will not be corrected but even may aggravate the subjective feeling of cramping. This foreign body encumbering the sternum, if too large and heavy in weight, will soon be condemned by the patient with the need of removal as a consequence.

However, custom-made silicone blocks are not sufficiently effective for highly deformed funnel chests and cannot correct the underlying structural defect with its associated functional problems. This means that this procedure leaves the physical condition absolutely unimproved, but may even deteriorate it due to psychologically triggered feeling of cramping by the weight burden of the implant upon the sternal region. The simple aesthetical correction of an extensive funnel chest without consideration of a functional rectification on the other hand may leave a problematic situation for these particular patients.

6.5.2 Fabrication of the implant

For the manufacture of the custom-made silicon implant a precise mold of the funnel depression must be made. For that purpose a special kit (Moulage Kit; McGhan Medical Corporation™, Santa Barbara, CA, USA; Fig. 9) is available. The mold alternatively may be made of different kinds of materials, like dental alginate [7], wax or plaster of Paris. This mold is manufactured in the surgeon's outpatient office, with the

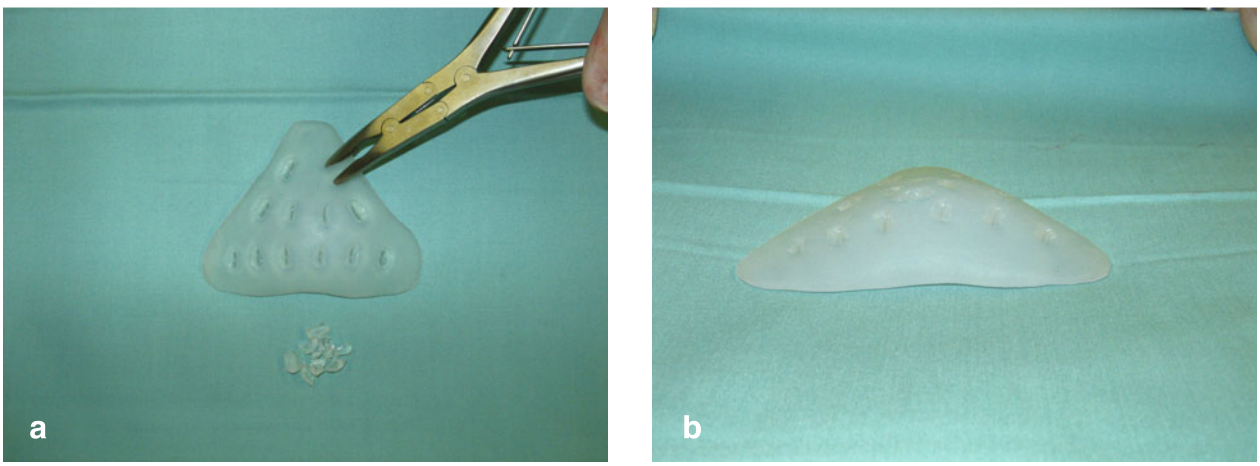

Fig. 6a. Custom-made implant during surgery, cutting holes into the implant to allow tissue ingrowths for stable and permanent fixation to the surrounding tissue. **b** Oblique aspect of the prepared smooth and textured silicone implant

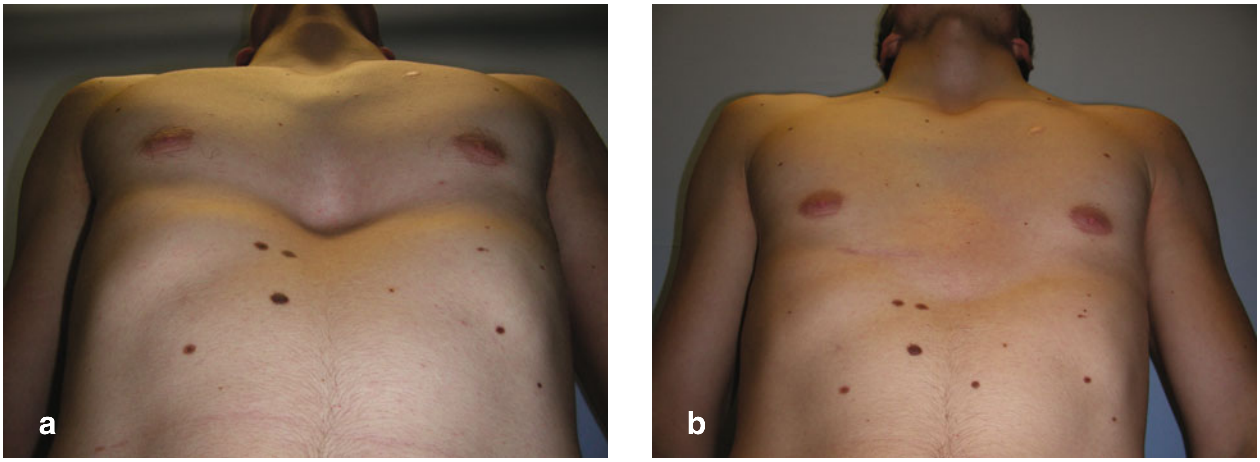

Fig. 7a. Frontal oblique view of a 23 years old man with symmetrical pectus excavatum. **b** Result of the same patient 6 months postoperatively

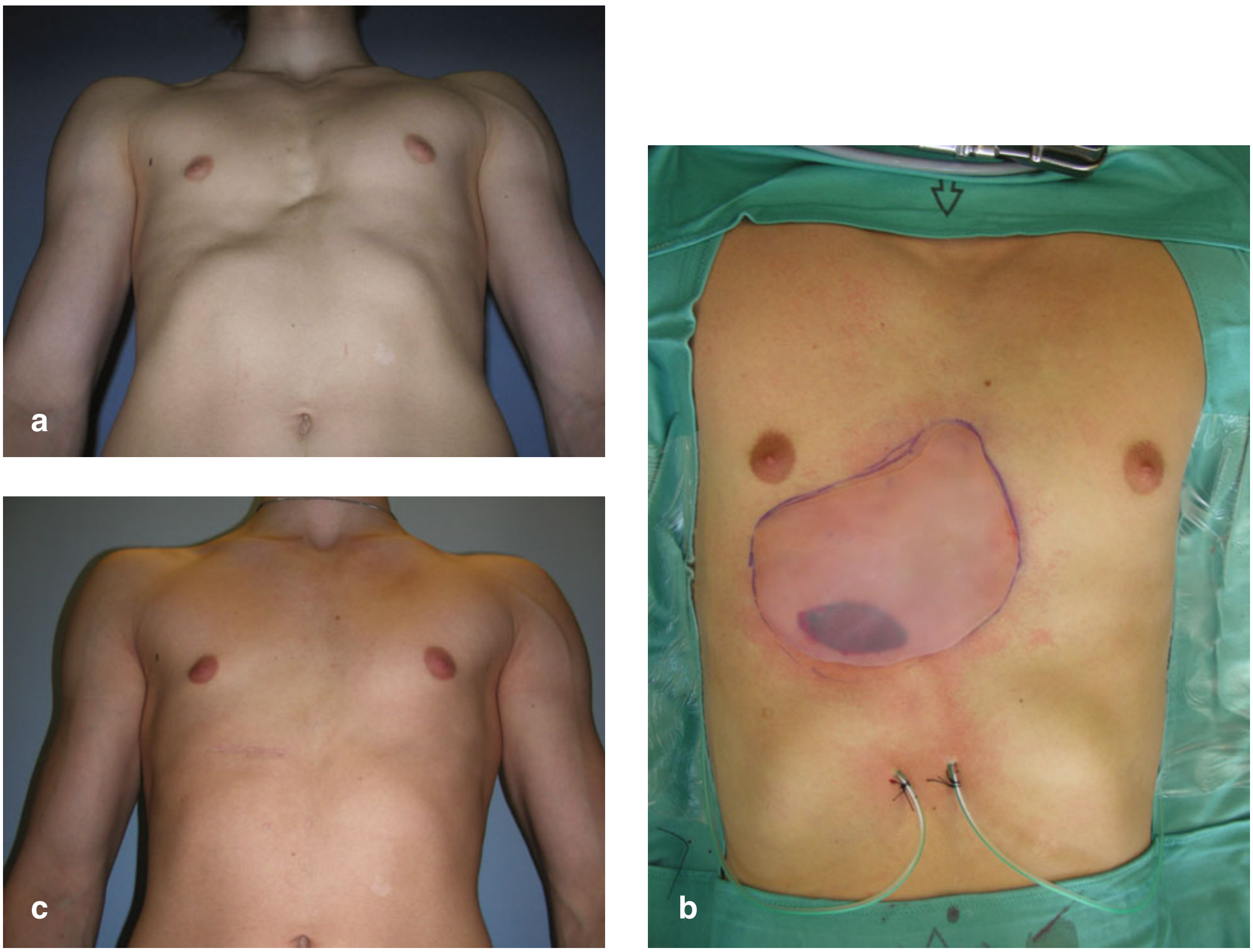

Fig. 8a. Preoperative situs in a 16 years old athletic male with asymmetry, markedly at the left side. **b** Intraoperative situs with the implant placed above the already dissected pocket. Notice the small skin incision behind the implant, sufficiently large for the passage of the folded implant and placed horizontally along the RSTL. **c** Situation 1 year after surgery with high contentedness of the patient

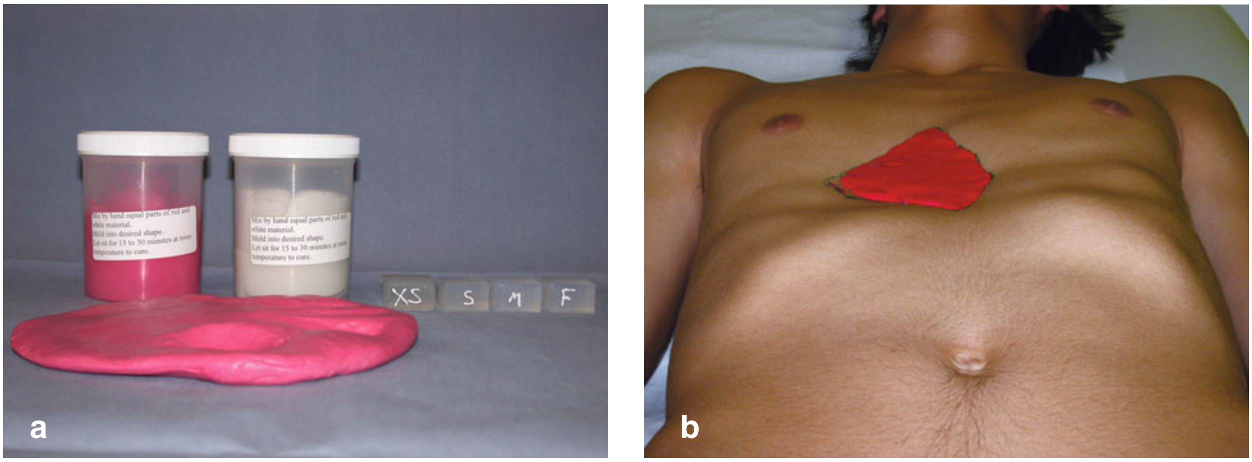

Fig. 9a. Exemplary moulage kit with the two components to be mixed manually. Depending on the exact percentage of the two components the vulcanizing time may last between few to many minutes, which allows secondary corrections after application to the patient's body. At the right side cube samples of available different softness qualities are depicted. XS, extra soft; S, soft; M, medium; F, firm. **b** Moulage adjusted but still soft in situ, waiting for vulcanization at room temperature into a firm template, simulating the extent of reconstruction already

patient in supine position (Fig. 10). Casting the implant, particular attention has to be drawn to the desired contour and especially to the edges of the mold to achieve a pleasant result of the corrected deformity without any undesired protrusion or rolling-up of its margins. Marks in 1984 [7] noticed that skin and subcutis thickness above the sternum differs from the thickness at the parasternal area, where it becomes thicker. This anatomical attribute also must be taken into account during subtle sizing the mold in order to prevent unpleasant contour deformity with an unnatural appearance after implantation of the final fabricate. It is also important to recognize that the skeletal defect may even be smaller than the external apparent defect, especially in well-trained athletic male patients (Figs. 7 and 8) or females with well developed or hyperplasic breasts. It is generally preferable to slightly undersize the mold in order to avoid protrusion and reconcilability of the whole implant or parts of it. Based on this mold and selecting the appropriate quality of softness or pliability a silicone implant will be fabricated (Figs. 9 and 12). In most of the cases the quality "soft" besides three others represents the ideal quality to fill up an uncomplicated excavation. Especially in slim patients or when the implant is subject of insertion via a remote skin incision, the quality "extra soft" might be advantageous for the passage through a long tissue channel.

Upon receipt of the final product from the manufacturer, noteworthy that the production processing may take several weeks, surgery will be scheduled.

6.5.3 Surgical technique

Surgery is performed with the patient under general or local anesthesia, the latter may be feasible in smaller defects only. In male patients, the skin incision (4–8 cm) is made horizontally in the direction of skin tension lines [10–12] for less visible scarring (Chapter 5.1), the length depending on the size and pliability of the implant (Figs. 10 and 11).

In female patients, an incision is preferably made along the medial part of the inframammary crease. A subcutaneous pocket is then prepared above the sternal periosteum and pectoralis major muscle fascia. Because of the elasticity/pliability of the silicone implant, a short skin incision sufficiently serves for insertion of a folded implant into the pocket (Fig. 12). For efficient blood coagulation in the subcutaneous pocket, a Langenbeck hook equipped with a light source likewise to an illuminated Biggs retractor [13] is used (Fig. 13). Afterward the silicon implant is inserted into the pocket and suction drains are left in place usually for at least 3–7 days, depending on the size of the pocket and extent of fluid production. To prevent postoperative hematoma, a compressive dressing with foam material is applied for 2–4 weeks. Dislocation of the implant during the postoperative period usually is no concern as long as the pocket matches the implant in size perfectly. That is why particular attention has to be drawn to proper dissection of the subcutaneous plane, exactly outflanking the borders of the implant and its designated

Fig. 10. Intraoperative situs of an athletic patient with an asymmetric pectus excavatum deformity. Notice the skin incision along the RSTL, the very pliable but large-sized implant and two drains already placed with a remote exit through the skin in order to reduce the risk of infection

Fig. 11. Skin incision camouflaged by setting it along the RSTL and into the hairy pectoral area

place. Otherwise dislocation especially along the gravitational forces to caudal or malrotation will become a cumbersome complication.

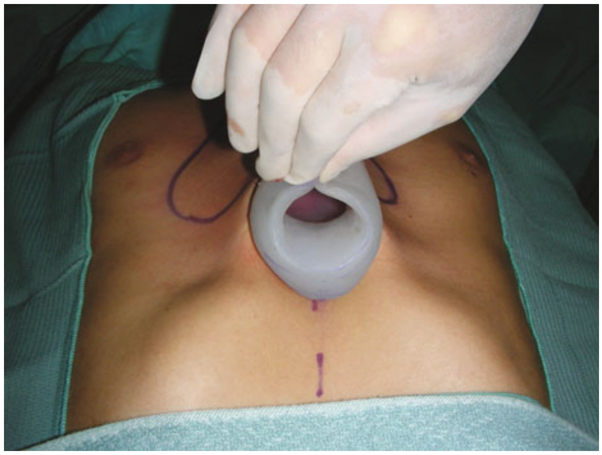

Fig. 12. Folding of the smooth silicone implant in order to keep the skin incision small

Fig. 13. A Langenbeck hook specially designed with a fixed plug to hook a cold light source in order to directly visualize structures deep within a tissue pocket

While some surgeons propagate a so-called “overriding” of the pectus excavatum deformity by implantation of breast implants, which usually are designed for breast augmentation, this procedure is

succeeding only under special circumstances. If for example an asymptomatic and only minor and particularly symmetric depression of the thoracic wall exists, with hypoplastic breasts, an exclusive breast augmentation can conceal the funnel breast deformity with implants sufficiently (Chapter 9).
As an alternative, Rose and Lavey in 1982 reported a simultaneous correction of bilateral breast hypoplasia and pectus excavatum using a single-unit prosthesis, consisting of two gel-filled breast implants firmly attached to a central silicone rubber implant [5, 6]. The main advantage of this attachment seems to be that no abrasion between otherwise loosely inserted implants takes place, which reduces the problem of seroma or even scar formation around abraded particles, based on potential foreign body reactions (Chapter 12.3). This abrasive process between a solid silicone block and onlay breast implants, should they have not been implanted in separate tissue pockets or should they have perforated through their own pocket subcutaneously into a conjoined pocket, is accounted for early leakage or rupture of the breast implants. As a modification Marks [7, 13] advocates the placing of the funnel implant deep to the muscles and muscle fascia. The particular advantage herein consists not only in the smoothening of the otherwise occasionally visible transition zone between implant and surrounding tissue, but also in a separation of breast implants from a funnel chest implant by strongly structured muscular and fascial tissue. However, in the presternal region this means pectoralis muscle flaps elevation and transposition to the midline.

6.5.4 Complications

Possible complications with silicone implants generally are result from heavy weight, too hard or too smooth consistency of the implant, formation of a fibrous capsule with ensuing contraction, formation of seroma, downward-slippage due to gravitation and visibility of the implant as a whole or at the edges due to rolling-up, rocking, prosthetic extrusion, late bleeding, and infection [4, 7, 8, 13, 14]. Loss of an aimed natural contour may be a result of overcorrection by using a too large implant, inadvertently again staining the patient. Especially the xiphoid region features a three-dimensional region that might be obliterated by imperfect shaping of the mold. Thus the natural angle of the costal cartilages with the xiphoid should be maintained as an aesthetically important area [13]. For the case that maintenance is not possible because of the nature of the depression, it should be reconstructed with the implant itself.
The high incidence of reported [7–9, 13] hematomas perioperatively is associated with the small access for implantation of silicone blocks in general. Short skin incisions are desired to minimize the length and reconcilability of a scar, the nowadays available silicone implants show sufficient pliability to pass through very small incisions. On the other hand such a minimal access does not allow optimal hemostasis during dissection of the subcutaneous pocket. Such wide ranging subcutaneous dissection for a widespread and flat implant also endangers the blood circulation of the elevated skin, the center of this “tent-roof” might develop necrosis if dissected too close to the skin surface, predominantly in slender patients. Anyhow, the implantation of such foreign materials in slim patients is a problematic endeavor, since almost every implant will be visible and appear in outlines through the thin skin, known as “implant show”. Therefore such an artificially created stain, that replaces the congenital depression indeed, will hardly satisfy the desires and requirements of an afflicted patient. Such a problem of implant show is predominately present in slim patients, but slender young adolescents and adults usually lack of transplantable fat too. Herein also the new advent of lipofilling will hardly fulfill the requirements of tissue augmentation in the presternal area (Chapter 6.6.2). Snel in 2009 [9] advocated the lipofilling procedure (autologous fat from abdomen, buttocks, thighs) several months prior to silicone implantation in order to enhance the thickness of the subcutaneous layer in the presternal area to avoid later implant show. An appropriate patient therefore must be properly evaluated for suitable fat deposits, sufficient in location and volume for serial lipofilling procedures. Even the microvascular transplantation of tissue flaps (Chapter 6.6.4) in such slender patients will probably not lead to a pleasant result, creating additional scars and show of flap contour beneath a very thin skin layer instead. Not even autologous transplantation cartilage chips, taken from occasional overgrowth of lower ribs (Chapter 6.6.1) might be helpful in such cases, because the agglomeration of these cartilage fragments will be easily palpable and worse, will be visible through the flimsy skin. In these cases it might be a wise solution to accurately inform and convince the patient to await maturation of the body for several years prior to perform surgery with an expectable unsatisfying result. In the meanwhile Macrolane™, a biological substance consisting of hyaluronic acid, may serve for temporary augmentation of the depressed area. Though very expensive so far,

multiply applied as a filler for facial wrinkles, this substance nowadays is utilized in breast augmentation too [15]. Yet without evidence-based studies and without available literature for the application in the funnel chest deformity the augmentation effect of this substance is reported to last up to 18 months in facial rejuvenation [16]. Due to resorption, which is an effect of natural degradation of its polysaccharide components, the augmentation or filling effect will vanish by time, thus the procedure must be repeated until body maturation will provide with sufficient transplantable subcutaneous fat tissue.

Besides hematoma formation seroma production predominately is the main concern in the utilization of silicone implants for reconstruction of the pectus excavatum depression. In patients in whom seroma occur, this effect may be caused by foreign body reaction to a widespread silicone surface (Chapter 12.3). This happening is quite unpleasant for the patient because serial aspiration is required in order to reduce pain and prevent over-distension of the skin with ensuing probability of remaining skin wrinkles, furthermore with every aspiration the danger of iatrogenic infection increases. However, aspiration of seroma is a requirement in order to avoid distortion or even dislocation of an implant floating within the seroma deposit.

Several authors [8, 9, 17] described 31–65% of patients developing seroma with the repeated need for aspiration due to undue skin bulging and pain. This phenomenon was described also by other authors [4, 7, 14]. However, in the earlier cases the utilization of different kinds of polymerized silicone was accused to that complication, but also nowadays, with the utilization of prefabricated and smooth textured implants this problem is still present. Thus informed consent must include this still very frequent complication. Even 20% of the collective treated in this series developed hematoma requiring secondary surgery for evacuation. The main cause for the high incidence of hematoma may be based on the transection of many intercostal and parasternal musculocutaneous perforating vessels, which immediately retract into the musculature and therefore are hardly accessible for proper hemostasis through a desirably small skin incision, the more, if this incision is placed remote from the implantation site for aesthetical reasons. Therefore the above-mentioned Langenbeck light-hook (Fig. 13) with the spotlight reaching into the deepest recesses is strongly recommended for the survey of a meticulous dissection and hemostasis. As an additional step against seroma or hematoma formation the application of an elastic, circumferential dressing, with steel- or cotton-wool serves with slight compression at the area of the implant. The compression must be adapted to the situation, that the elevated skin flaps, due to transection of many musculocutaneous perfora-

Fig. 14. Implant with a textured surface. This kind of superficial structure apart from material tongues or incisions also is suitable to prevent dislocation and should enable firm attachment to the surrounding tissue.

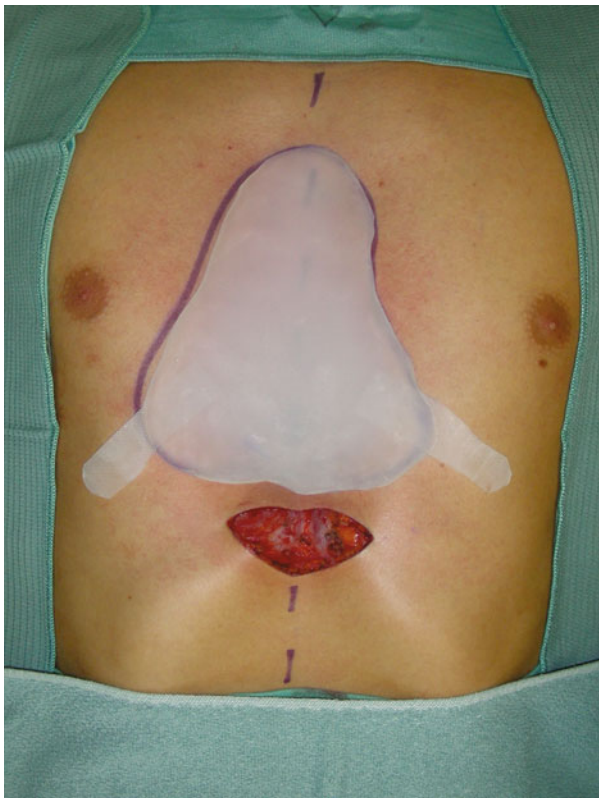

Fig. 15. A large smooth silicone implant equipped with silicone straps, designed for additional subcutaneous fixation to prevent rotation

tor vessels possess lesser perfusion, thus the development of skin necrosis can be a result of too tight dressing.

According to the report of Marks in 2000 [13], but currently still with lacking evidence due to the available small cohorts of patients, seroma formation is decreased when the implant is fabricated with a textured surface (Fig. 14). This texture additionally allows tissue ingrowths into the rough superficial structure thus provides with firm adherence and subsequently decreases the risk of displacement or rotation as well as seroma formation. Another possibility to prevent rotation is the fabrication of silicone straps fixed to it. Those straps may be adhered to fascial structures with non-absorbable sutures (Fig. 15).

Asymmetric deformities of the anterior thoracic wall with tissue depression, not diagnosed as funnel chest, but summarized as miscellaneous deformities (Chapter 2.3), appearing as a single deformity or conjoined with other deformities may also be subject of treatment with silicone implants [18]. Particularly lateral depressions of rib cartilages and bones are hardly accessible for any type of surgery, as well as depressions at the lower thoracic wall involving the seventh to tenth rib. Besides congenital deformities, asymmetric flattened or even caved deformities may also be an iatrogenic result of remodeling an asymmetric funnel chest. Even in the keel chest deformity such excavations may be present and cannot be resolved by conventional rib and cartilage remodeling. For such cases autologous tissue augmentation like lipofilling (Chapter 6.6.2) or cartilage chips (Chapter 6.6.1) may serve for permanent improvement, but also custom-made silicone is ideally suitable for these minor, due to predominately asymmetric appearance and thus aesthetically unpleasant defects (Figs. 16a, b and 17a–c).

Molding and prefabrication does not differ from the technique described above. Placing of the incision along the RSTL (relaxing skin tension lines) may be remote from the midline or the defect itself in a hidden area. Usually such depressions present shallow but widespread, so that a prefabricated silicone implant may easily be folded or rolled for insertion through a minimal skin incision. The reluctance against such a small port for implant insertion is the limited survey during pocket dissection. This problem may be overcome either by using an illuminated skin retractor (Fig. 13) or an endoscope for the case of a very distant skin incision [19, 20].

Overall, the silicone implant in the hands of experienced surgeons shows contentedness from 69% up to 95% of the patients treated even in the long-term evaluation [8, 9]. Thus and because major surgery can be avoided, this alternative can serve as a first choice of treatment in selected cases for aesthetic

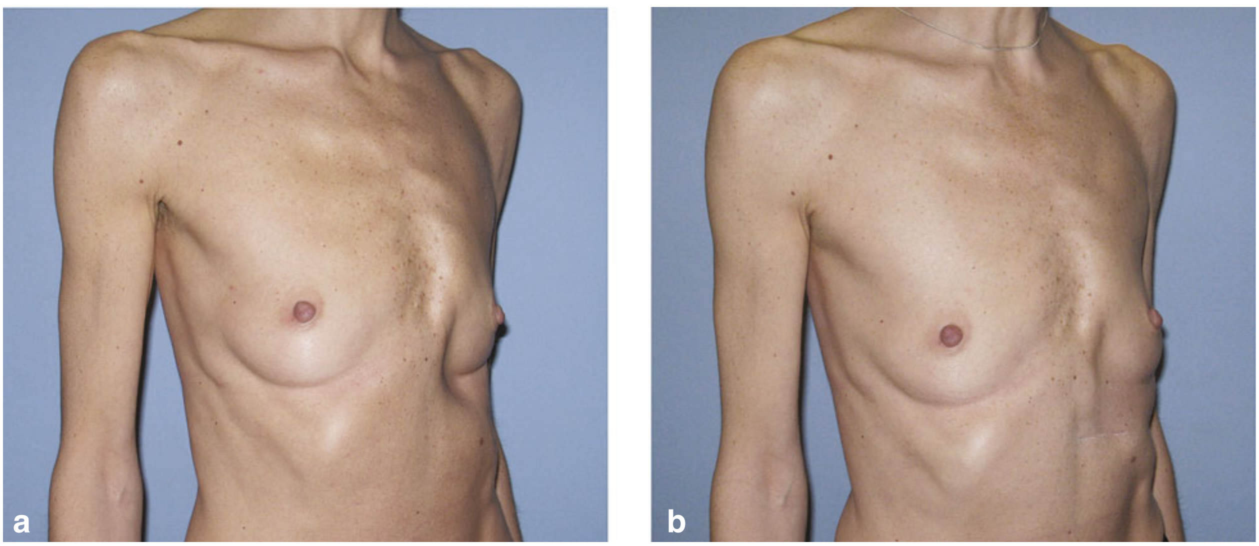

Fig. 16a. A 45-year-old extremely slim female with asymmetric depression at the left lower thoracic region, distortion of the left breast to caudal and inaccessible for distinct thoracic remodeling surgery. **b** Same patient 14 months after implantation of a flat and soft custom-made silicone "leaf" with stable and aesthetically very satisfying result. Even the breast on the left side improved its projection now almost symmetrical to the contralateral unaffected side

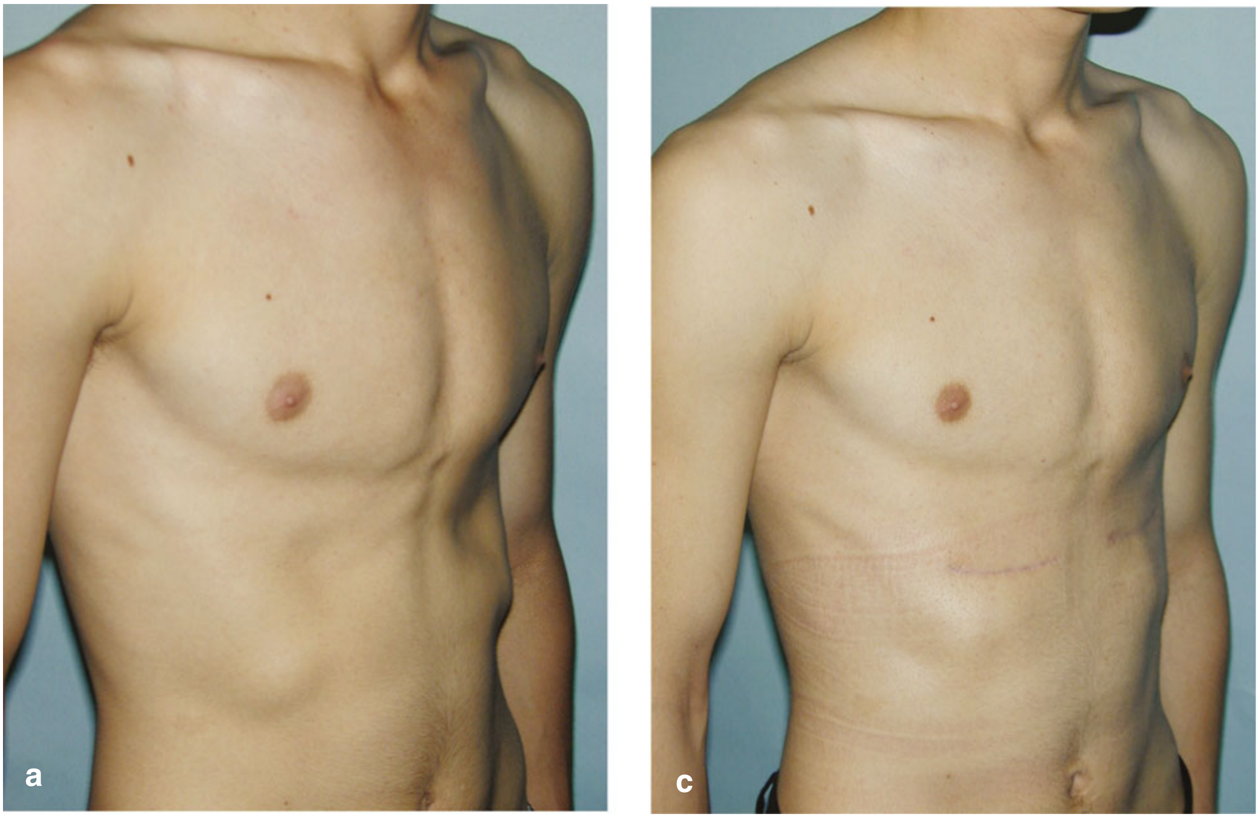

Fig. 17a. A 18 years old male preoperatively with major depression of the ribs at the left infrapectoral region and minor at the right side. **b** Custom-made silicone implant with the quality "XS" (extra soft) upon receipt by manufacturer and prior to sterilization. **c** Same patient 6 months after subcutaneous silicone augmentation

improvement of the pectus excavatum deformity, despite the high incidence of minor complications. Silicone implantation is ideal in adults, when the body has reached its final skeletal shape and contour and cardiopulmonal impairment is absent. Nevertheless many patients refuse the use of foreign body, but alternatives with transplantation of autologous tissue in many cases can afford relief of such circumstances, however, creating donor defects and additional scarring. Informed consent must contain all the advantages and disadvantages of silicone and alternative procedures as well [9].

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6.6 Autologous tissue

6.6.1 Cartilage chips for treatment or refinement in funnel chest deformity

Anton H. Schwabegger

The application of autologous cartilage is well established exemplarily for nose reconstructions or other regions at the face already since many years [1–7, 9]. Such autologous cartilage grafts, herewith harvested from deformed ribs preset themselves also very well suitable to fill up minor or minimal funnel chest deformities. In contrast to alloplastic material, such as silicone or artificial bone, autologous grafts serve as volume replacement lifelong, as far as they grow in without resorption. They exemplarily can be used for the adjustment of asymmetrical contours and small remaining irregularities, which still annoy the patients after corrections of a funnel depression using a pectus-bar or applying other techniques. Sometimes and not before several months after the elevation of an asymmetrically one-sided distinct deformity, an undesirable gibbus at the contralateral side then may result as a minor but well-recognizable hump. This gibbus, it usually concerns only single rib cartilages directly at the junction zone to the sternum, can be resected out or cut tangentially within the same surgery as the bar explantation, however using separate incisions presternally or parasternally. These cartilage parts as a by-product may then be used to fill up remaining small-depressed areas of the anterior thoracic wall, which were not manageable by the initial surgery. In contrast to the application in nose augmentation, the rib grafts are thoroughly cut to small chips of not more than 5 mm in diameter by scalpel, wrapped into bioabsorbable hemostyptic and antimicrobial fabric (Tabotamp™ or Surgicel™ by Johnson & Johnson Corp., New Brunswick, NJ, USA) and then placed into a subcutaneous pocket encompassing the concerning depression (Fig. 1a–g).

If such an asymmetrical contour with unilateral depression and projection of a gibbus should become apparent immediately during implantation of a pectus-bar with a semi-open access, when cartilage rib parts devolve from necessary chondrotomies, the resulting contour deformity may be adjusted to a symmetrical surface immediately utilizing these cartilage chips (Fig. 2a–c).

In minor but distinct forms of the pectus arcuatum deformity such cartilage chips-transplantations outstandingly suit for correction, if for example osteotomies or a pectus-bar implantation meant surgical over-treatment and solely the necessity consists to cut off excess of those cartilages which cause the gibbus (Fig. 1a–e, Chapter 8).

The cartilage pieces to be resected are either taken subperichondrially or otherwise meticulously freed from perichondrium. The cleansing of that perichondrium is mandatory, because reimplanted perichondrium could, above all at the youth, which are subject of skeletal growing, produce undesirable contour irregularity through cartilage neogenesis and therewith even might deteriorate the intended aesthetic improvement and final result.

The cartilage chips become minced to small pieces of not more than 5 mm in diameter (Fig. 1a) using a Lür, a Liston or preferably scalpel. On the one hand this diminishing of the cartilage grafts to small chips has the advantage that this cartilage gravel can easily be contoured after the implantation into the subcutaneous tissue pocket (Fig. 1f). So that however this cartilage gravel does not arrive at undesirable localizations, it is enveloped with rapidly bioabsorbable cellulose fabric layers for example, is moistened with saline (Fig. 1c) thus temporarily fixed in a desired shape [4, 8]. In addition or instead of this cellulose fabric, also Spongostan™ (Johnson & Johnson Corp., New Brunswick, NJ, USA) or fibrin glue (Tisseel™, Baxter International Inc., Deerfield, IL, USA) can be used [3, 8]. Ideal shaping of the cartilage gravel though is dependent on an exactly dissected tissue pocket, and for the case of embedding with fibrin glue, after its hardening that occurs within seconds, adaptive shaping is hardly possible any more. That means that prior to the injection of fibrin glue the shaping or modeling of the cartilage gravel must be terminated already.

On the other hand, mincing of the cartilages augments their surface to establish sufficient contact with surrounding vascularized tissue, which enables ingrowth and prevents necrotization with ensuing resorption.

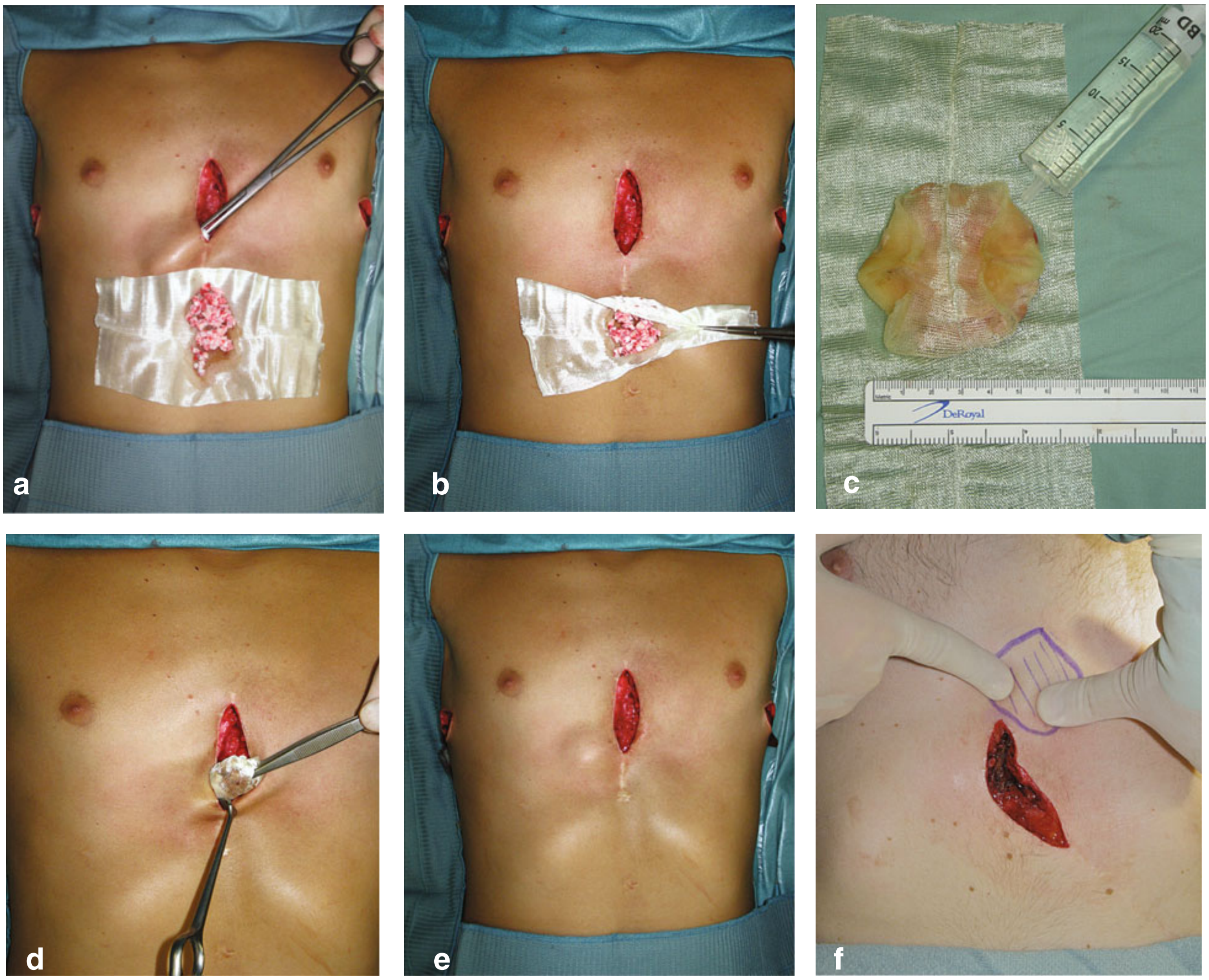

Fig. 1a. A 16-year-old boy with pectus excavatum, corrected by MIRPE and minimal anterior access for supportive sternum elevation through an old presternal scar. Simultaneous augmentation of a right-sided depression (forceps) with autologous cartilage chips already cut and placed on a Surgicel-sheet. **b** Wrapping of the cartilage grafts with Surgicel. **c** Moistening of Surgicel for smooth package of the cartilage grafts. **d** Implantation of packed grafts into the dissected subcutaneous pocket. **e** Implanted graft mass, yet causing exceeding hump, prior to molding. **f** Manual molding of subcutaneously (marked area) implanted grafts to obtain a plain and inconspicuous skin surface, demonstrated in another 21-year-old patient with simultaneous correction of an old scar, resulting from diaphragm surgery in childhood. **g** Final situation after molding (patient of e) and prior to wound closure

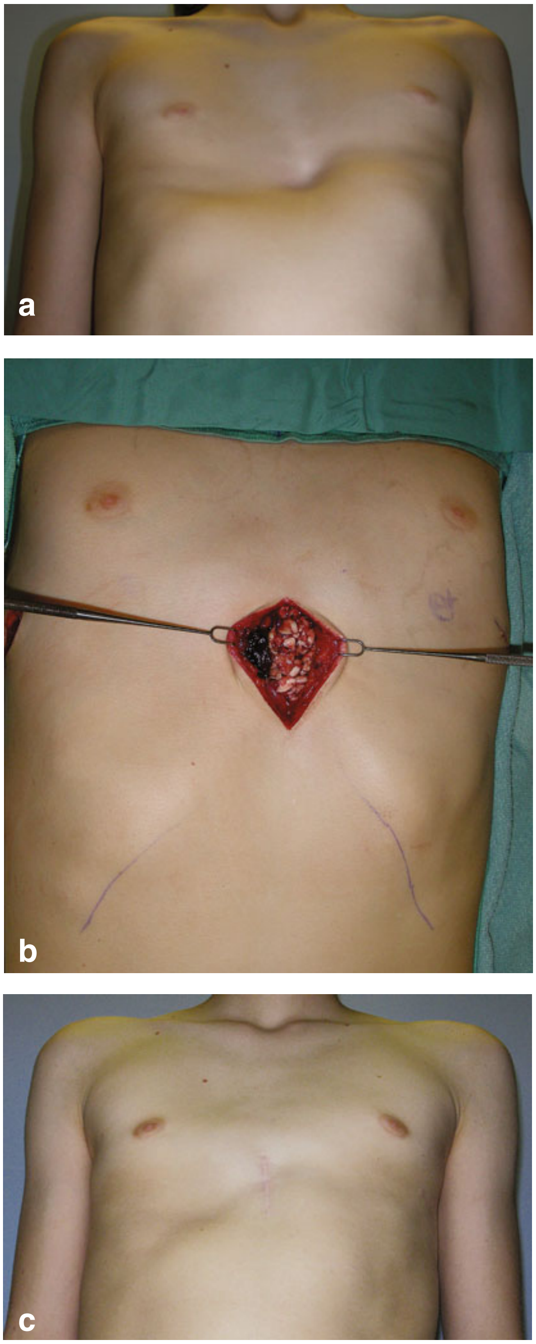

Fig. 2a. A 16-year-old male with asymmetric pectus excavatum, prior to MIRPE and minimal central access. **b** The MIRPE procedure caused gibbus formation at the left side, the resulting hump was simultaneously resected and the by-product cartilages were implanted as chips to egalize the remaining irregular depression for aesthetic improvement of the skin surface. **c** Situation 1 year after pectus-bar explantation, 2 years after initial surgery and simultaneous cartilage grafting

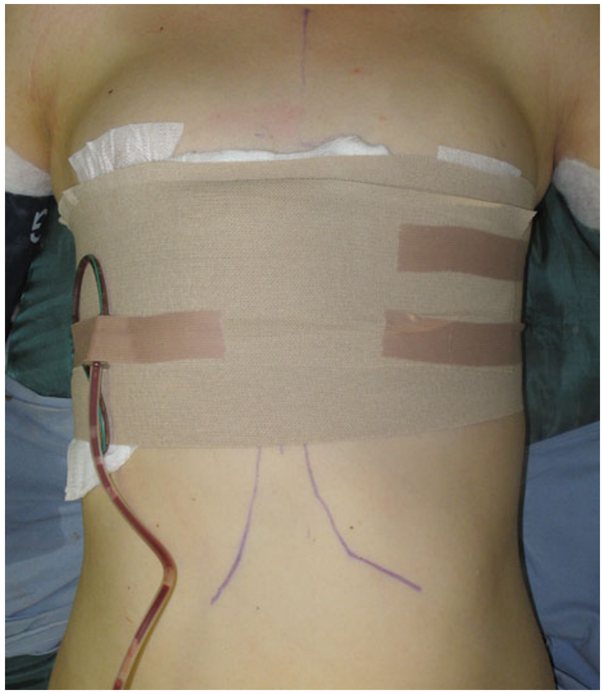

Fig. 3. Elastic bandage for moderate compression of transplanted autologous cartilage chips to avoid dislocation and to enable close contact of vascularized tissue for rapid ingrowth thus minor resorption

In contrast to foreign material like silicone implants (Chapter 6.5) autologous cartilage on a long-term basis is more durable and also more economical. However, the maximal layer thickness of the cartilage gravel should not exceed 1 cm so that a complete ingrowth of the cartilage chips with connective tissue and scar is enabled and therefore disintegration of transplanted tissue by resorption processes caused by lesser or lacking perfusion can be avoided. This maximal layer thickness of 1 cm therefore means the (personally experienced, no valid data available so far) limiting factor for the filling-up of a funnel chest depression in one stage. Partial resorption can be disadvantageous, which under circumstances may produce contour irregularity of the engrafted mass. This unpredictable irregular resorption rate still means a major issue in cartilage grafting, published in series of reconstruction or augmentation rhinoplasty [1]. However, such irregularities at the nose will much more bother a patient due to aesthetic thus psychological reasons, but will not at that extent in the patient with pectus deformities. That is because the presternal and surrounding skin presents much more rigidity than the skin at the nose and therefore

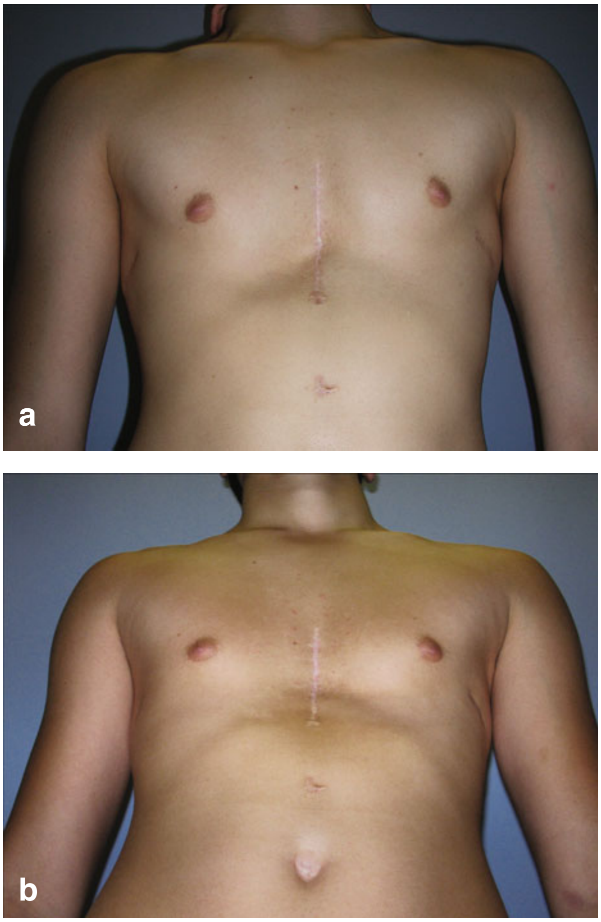

Fig. 4a. A 17-year-old boy, situation 1 year after pectus-bar implantation according to the Nuss procedure, combined with minor open presternal access, prior to pectus-bar explantation. Notice remaining asymmetrical depression at the left paraxiphoid area. **b** Same patient 11 months after pectus-bar explantation and simultaneous depression augmentation with cartilage chips, resulting in pleasant contour symmetry

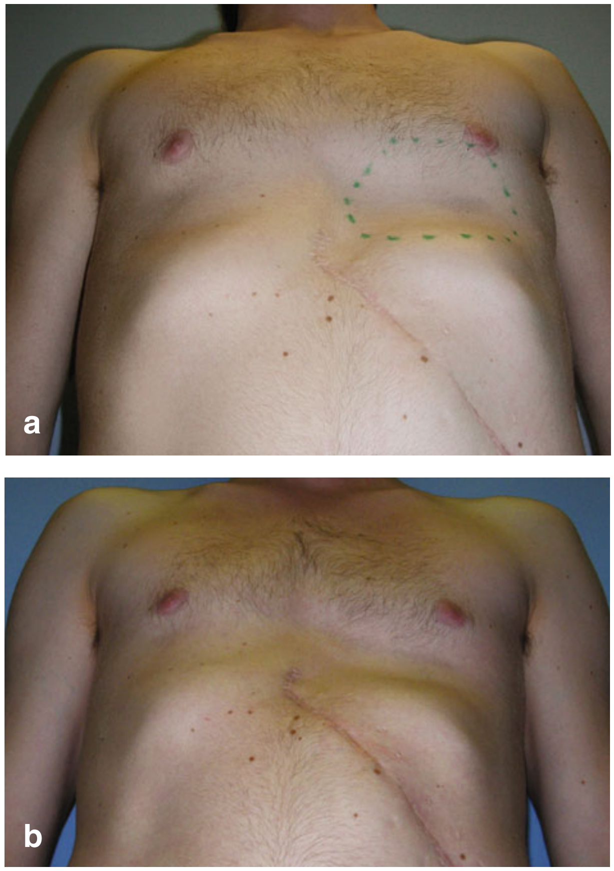

Fig. 5a. Same patient as in Fig. 1f, situation 3 years after MIRPE with one bar, prior to its explantation, notice circumscribed (marked) rib depression at the left side. **b** Situation 1 year after pectus-bar explantation and sufficient improvement of the left-sided depression by simultaneous augmentation with autologous cartilage chips

irregularities are much better concealed by that quality. Furthermore, slight surface irregularities resulting from partial resorption of grafts in most of the cases still cause contentedness of the patients because of mostly sufficient augmentation of any cavity. If augmentation of a deeper depression solely with cartilage chips is aimed, it should be scheduled and performed in two or more stages, separated by intervals of at least 3 months. The encompassing area of tissue augmentation on the other hand is limited through the quantity of available cartilage that can be harvested without resulting problems at the donor site. Until the fixation of the transplanted cartilage gravel by endogenous embedding scars, a rib bandage (cingulum) with moderate compression onto the modeled region should be carried for 4 weeks (Fig. 3).

Above all in the secondary corrections with aesthetic refinements, the cartilages available by necessary resections usually suit outstandingly for contour adjustments of smaller asymmetric sunken defects caused by any prior remodeling procedure (Figs. 1a and 4a, b). On the other hand, especially in very slim patients, the contours of cartilage grafts placed subcutaneously may be visible and easily palpable. Furthermore in the primary intervention of only small or moderate defects, when no surplus of cartilage is available, the removal of cartilage out of "healthy" regions is not indicated or not desired. Therefore further methods are described in Chapter 6.6.2; exemplarily the nowadays re-developed and refined technique of lipofilling substantially appears more suitable in distinct cases because of its simplicity and lesser invasiveness, however, as long as sufficient subcutaneous fat is available for liposuction.

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6.6.2 Lipofilling for funnel chest and similar or adjacent anterior thoracic wall deformities

Monika Mattesich, Anton H. Schwabegger

6.6.2.1 Introduction

The transfer of autologous fat as whole grafts has been performed since the end of the 19th century and as injectable grafts since the 1920s; however, it is only within the past 20 years that the popularity of autologous fat transfer for the correction of various deformities has increased within the plastic surgery community. The rising interest in this procedure has paralleled the development of liposuction for body contouring. One reason may be the natural tendency to capitalize on the opportunity that the removed fat may be used to augment or restore areas at the body or face [17] with volume loss, contour irregularities or congenital deformities. Plastic surgeons most experienced with fat filling procedures have reported good clinical results suggesting short- and long-term persistence of transferred grafts, and thus promote autologous fat as the ideal filler [8, 10, 13, 22].

Despite the initial clinical optimism associated with autologous fat transfer, there remains an uncertainty among physicians regarding the viability of transferred fat [2]. A critical assessment of the literature seems to object to the consistent success of the procedure. Animal studies by Peer [18] have failed to demonstrate the longevity of transplanted fat. Furthermore, only few clinical reports have applied objective outcome measures that document fat survival. Surprisingly, this has not decreased physician's enthusiasm for this technique. Modifications of the original techniques have been reported by Coleman, who promotes an atraumatic method of fat harvesting, centrifugation, and reinjection with the aim of maximizing nutrition and structural integrity at the recipient site [4]. He reports high physician and patient satisfaction from his "structural fat grafting" technique [9]. The heightened interest in autologous fat transfer by patients and physicians should arouse objective investigation. Advances in three-dimensional surface mapping (see Chapter 9.1) with laser scanners and professional photography (see Chapter 3.1) now allow inexpensive methods for documentation of fat grafting techniques. It is assumed and hoped that future tissue engineering techniques with adult-derived stem cells will admit long-lasting results and more predictable outcomes [16].

6.6.2.2 History

Neuber at the 22nd Convention of the German Society of Surgeons gave the first lecture on autologous fat transplantation to the periorbital region in 1893. It were Lexer, Czerny and Rehn who reported on similar methods in thoracic-, abdominal- and breast-surgery over the following decades. In 1911 Bruning used autologous fat for the correction of subcutaneous defects [1].

Although there was initial optimism expressed in these reports, subsequent studies noted that grafts diminished in size after transplantation. These studies recommended overgrafting to account for the loss in restorative volume by natural resorption. Marchand histologically examined transplanted human fat tissue blocks and observed that a large central portion was nonviable after 10 weeks; however, peripheral sections bordering connective tissue showed proliferation. In his review of the literature in 1923, Neuhof reported that fat undergoes a similar process like transplanted bone, whereby fat dies and is replaced by fibrous tissue or newly formed fat.

In 1950, Peer documented that free fat grafts loose approximately 40% of their weight and mass per year. On the basis of microscopic assessments of harvested grafts, he assumed that durable fat cells were concentrated in the center core of a graft and the remainder of the cells degenerate. In a follow-up report Peer emphasized the importance of both the need of hemostasis to maximize survival of the graft and a good blood supply at the recipient site [19].

These findings were similar to those of other investigators over the subsequent three decades [16]. This corresponded to a declining interest in fat grafting by physicians who were concerned about inconsistent and unpredictable results and therefore the technique was not widely used.

6.6.2.3 Current investigations

In the early 1980s, liposuction turned out to be the most frequently performed procedure of aesthetic surgery and it provided plastic surgeons with a valuable by-product: semi-liquid fat that could be grafted with relative ease using needles or small cannulas [9].
In many ways, fat is the closest that exists to an ideal filler: it is readily available and not expensive; it is autologous and therefore lacks a host immune response; it is safe and noncarcinogenic; and it is easily acquired with a minimal invasive procedure [15].
In the early 1990s, a series of positive reports of fat grafting were published [11]. One of the promoters among them was Sydney Coleman, who postulated that "his experiences with fat grafting have confirmed the efficacy and permanence of grafted fat, provided that it is handled atraumatically and that proper harvesting and grafting technique is followed". His consistent publications show that fat grafting can result in long-term corrections [3–7]. He began to observe other attributes of fat. He noted that the quality of the skin under which grafted fat was placed improved, not only as an effect of the fullness but also with gradual improvement in the quality of the skin. Wrinkles softened, pore size decreased, and pigmentation improved over the first year. There also appeared to be an improvement in the quality of the tissues into which fat was grafted. In the late 1990s, he reported that fat grafted under depressed scars not only relieved the depression but also seemed to soften or even completely eliminate the visibility of a scar, turning the appearance to look like normal skin, so that the effect of grafted fat seemed to be much more than just a long-term filling [5].

6.6.2.4 Indications

Nowadays, the technique of autologous fat transfer is widely spread. We can find numerous publications on its applications for all kinds of subcutaneous soft-tissue defects, lipoatrophies [17], the treatment of breast capsular contracture [24], chronic skin ulceration [12], the treatment of radiation skin damage [14], of mild pectus excavatum [20] and its application in Poland's syndrome [23].
In this chapter the emphasis is put on the one hand on the application of autologous fat transfer for the correction of mild chest wall deformities as a primary therapy option. On the other hand, it has shown to be a valuable alternative for the correction of residual defects of chest wall deformities after minimally invasive, open or combined correction techniques (Nuss, Ravitch, etc.) and after correction with custom-made silicone chest wall implants or conventional breast implants, respectively.
It can also be taken into consideration for the aesthetic correction of distinct presternal scarring.

6.6.2.5 Technique

The procedure is usually performed under general anesthesia. Patient's position is supine; fat is taken from the abdomen or thighs in the conventional wet technique. At our department, we use a special liposuction device (Tissu-Trans® by Shippert Medical Technologies Corporation, USA). Fat is harvested in routine fashion using back and forth motion. It will remain in the specially designed syringe with a multitude of sidewise perforations, and all blood, fluid and waste will be suctioned out to the waist canister (Figs. 1–4).
When the syringe is full or the desired amount of fat has been obtained, the harvest tubing is removed from the outer casing, a sleeve is pushed over the fat-filled syringe and a plunger is inserted into the end of it. A cannula is attached and the harvested fat is now ready to be re-injected into the designated areas.

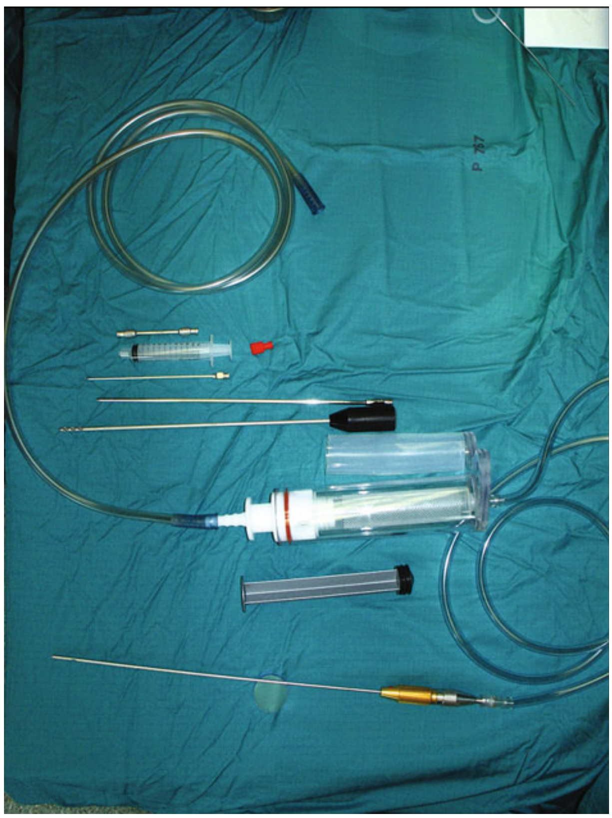

Fig. 1. The Shippert Tissue-Trans™ Model 60 for liposuction and simultaneous reinjection utilizing the same 60 ml syringe

Fig. 2. Technique of liposuction exemplarily at the abdomen to harvest an appropriate amount of fat tissue for transplantation

Fig. 3. Occasional fat separation from dispensable fluid and transfer to small syringes

After successful suctioning, local anesthesia with adrenalin as a vasoconstrictor is injected into the donor site to avoid hematoma.

6.6.2.6 Graft placement

Fat grafts are placed into the subcutaneous layer. Injection is undertaken via a fine cannula with multiple passes, injecting only tiny amounts with each pass and with deposition of fat in chains as the cannula is withdrawn. Ideally, fat is deposited at varying levels in a fan-shaped and atraumatical manner as this increases the surface area for contact with nourishing capillaries, as well as providing a more desirable aesthetic result (Fig. 5).

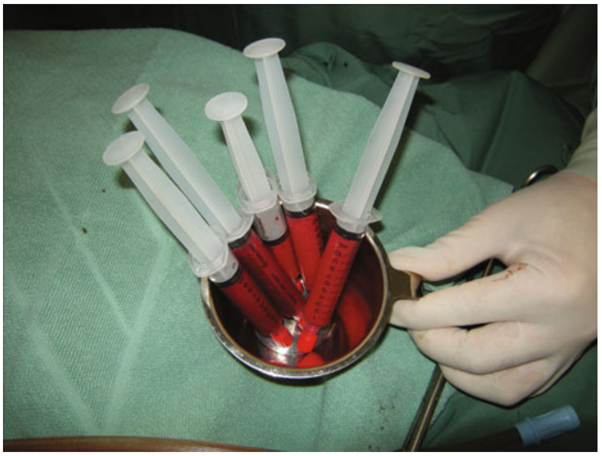

Fig. 4. Fat transferred to small syringes for a more convenient handling and portioning during injection

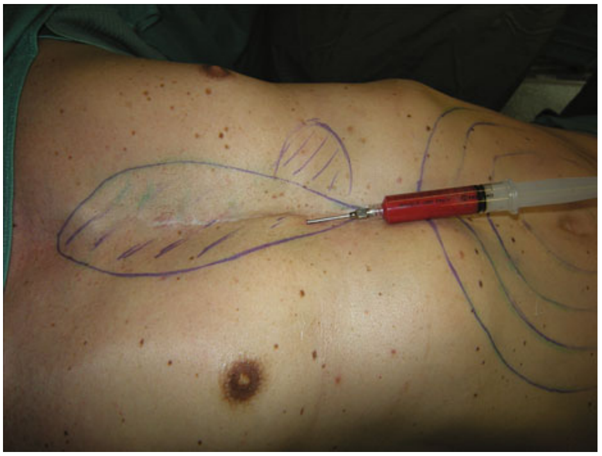

Fig. 5. Technique of injection of fat tissue in chains, multiple layers and fan-shaped, in order to prevent resorption

6.6.2.7 Discussion

Although there is a current trend in replacement and recontouring of almost all kinds of subcutaneous tissue deficits with autologous fat transplantation [25], the literature fails to provide definitive evidence of survival of the transplanted fat [26]. Modern molecular biology techniques such as cell-labeling and the ability to monitor cell trafficking will provide more answers to clinicians' questions. In addition, a large-scale clinical assessment using three-dimensional volumetric imaging would supply useful data (see Chapter 9.2).
Nevertheless, several but so far preliminary results indicate that lipofilling can improve aesthetic results after correction of various chest wall deformities in-

cluding the female breast. It is speculated that this improvement depends on tissue regeneration promoted by adipose tissue-derived stem cells. However, further research must be undertaken to assess these adipose cell properties, extra-cellular matrix composition, and the essential requisites for routine clinical applications should be investigated prior to a routine application.

The morbidity related to lipofilling is rather minimal, similar to that for a limited liposuction, with acceptable safety. It is a reliable and reproducible technique with negligible residual scarring and few complications (such as fat necrosis). In order to reduce the risk of fat necrosis, a strict injection technique should be adhered to and excessive injection of fat should be avoided. It is virtually impossible to define an ideal volume that should be transplanted, because this depends on various factors such as patient's morphology, the amount of fat available, the quality of the skin and the volumes and location treated. Injection of a maximum amount of 200 cc per session is recommended, as long as the technique of fan-shaped injections into different layers is feasible.

Promising results documented for other applications of lipofilling give hope for equally positive outcomes of its application in minor chest wall deformities, for the adjustment of asymmetrical contours and remaining small irregularities that might still annoy patients after various corrections as the implantation of a pectus-bar or other techniques (Figs. 6, 7).

Fig. 6. A 46-year-old man with recurrent funnel chest deformity after several operations elsewhere (Rehbein approach in childhood, Ravitch and pectus-bar technique as an adult). For a further aesthetic improvement and treatment of the adherent presternal scar one session of lipofilling with 100 cc was performed

Fig. 7. Preliminary result 2 weeks after lipofilling for release of the presternal scar and minor tissue augmentation. A second and possibly third session of lipofilling may continue the aesthetic improvement

An autologous fat tissue graft, mostly used as a complementary treatment, improves trophicity if the integument is thin, gives better definition to the reconstructed contours and adds bulk in completion of the

Fig. 8. A 42-year-old woman with persistent asymmetric pectus arcuatum deformity after several operations with breast implants on both sides and sequential presternal custom-made silicon elastomer implants. Lipofilling with 55 cc to the left breast and to the presternal region. Simultaneous correction of the ipsilateral submammary fold and of a remaining bulk of the second rib cartilage on the right side

Fig. 9. Preliminary result 6 weeks after lipofilling. Minor improvement at the funnel deformity itself but notably better contouring at the inner and upper margin of the left breast implant. The remaining funnel would need further lipofilling

conventional techniques. However, several injection sessions are necessary due to the unpredictable amount of resorption of the transplanted tissue, but can often be associated with other corrective or refinement procedures.

Lipofilling enables improvement of the quality of the results, which often seem to be imperfect if only a single approach or operation technique is performed.

In the future autologous fat injection for minor chest deformities may be considered as an alternative to other more invasive procedures. Lipofilling might replace the implantation of small custom-made silicone elastomer implants and can even be applied for larger presternal depressions. It is also applicable for the correction of breast asymmetries associated with the thoracic wall deformity (Figs. 8, 9), since the radiographic appearance after injection does not seem to lead to confusion with malignancy and does not interfere with such diagnostics so far [21].

It should be kept in mind that the discomfort of minor chest wall deformities is almost always aesthetic and that in most cases it has no functional impact. Surgeons must therefore propose the most appropriate treatment for each patient, causing of course the least morbidity, leaving minimal scarring and functional sequelae, which may be credited as an argument for autologous fat transplantation by the lipofilling technique in minor and moderately sized deformities or as an adjunct to refine major corrections.

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6.6.3 Local flaps

Anton H. Schwabegger

If invasive methods of thoracoplasty like sternum turn-over, costochondrosternal remodeling or less invasive MIRPE surgery are refused by the patient or contraindicated due to normal fitness and/or only minor expression of the pectus excavatum deformity, local or distant transposition of autologous tissue, may be an alternative treatment option. However, generally in children and adolescents as well as in adult males never sufficient tissue volume is available in the parasternal region for the purpose of filling up a funnel depression through simple tissue transposition. Distant flaps like the TRAM-flap [9] or an omentum-flap [3] exemplarily can be transposed into a central funnel chest defect indeed, in analogy to defect cover for example in osteomyelitis of the sternum or after tumor resection [2, 15]. Even a vascularized rib strut [7] may aid in remodeling, but therein is used as a living support for the elevated sternum, that is mobilized according to the standard Ravitch procedure. Thus such a vascularized rib strut will not directly contribute to volume augmentation, but rather will maintain the position of an elevated sternum.

In predominantly aesthetic indications and young patients, such invasive interventions interfering with the body static or with opening of the abdominal cavity are considered rather contraindicated, however, as long as simpler and less invasive methods are available. Alternatively a pedicled latissimus dorsi muscle (LDM)-flap may be transposed [14], but with the disadvantage of loosing the posterior axillary fold and muscle function to some extent [1]. Moreover to reach the funnel cavity sufficiently just by transposition, the LDM must be detached from the humerus and furthermore its (thoracodorsal) nerve must be transected to avoid inadvertent muscle contractions at the anterior thoracic wall. This denervation will cause atrophy of the muscle, which especially in slim male patients will result in reasonable loss of volume within the augmented space thus will lead to minor aesthetic outcome, if not overcorrected by inclusion of fibroadipose tissue in advance [14]. On the other hand, if transposed in youth and slenderness, transposed fibroadipose tissue adjacent to the LDM may become bulky with increasing body mass during maturation, thus resulting in an unpredictable tissue hump localized widespread above the sternum and parasternal area.

Free flap transfer as a further alternative may be a pains taking endeavor concerning microsurgery, if one is not experienced or routinely dealing with such procedures, but nevertheless can be very helpful in large-scaled pectus excavatum deformities (Chapter 6.6.4).

In adult women on the other hand, with eventually hyperplastic breasts or breasts grown asymmetrically (Fig. 1), the surplus tissue of one or both breasts can be used in place of a custom-made silicone implant (Fig. 2) in order to fill the presternal cavity. In analogy to a reduction mammoplasty according to the method after Morestin in 1907 [6] and Piza [8] a discoid-shaped part of breast tissue is dissected (Fig. 3) out of the concerned breast. Only singular reports are available in the current literature concerning the utilization of such regional tissue for the correction of a funnel deformity. Guimaraes in 2001 described the utilization of adipose tissue flaps [4],

Fig. 1. A 24-year-old female patient with moderate but asymptomatic funnel chest deformity and asymmetric breast hypertrophy

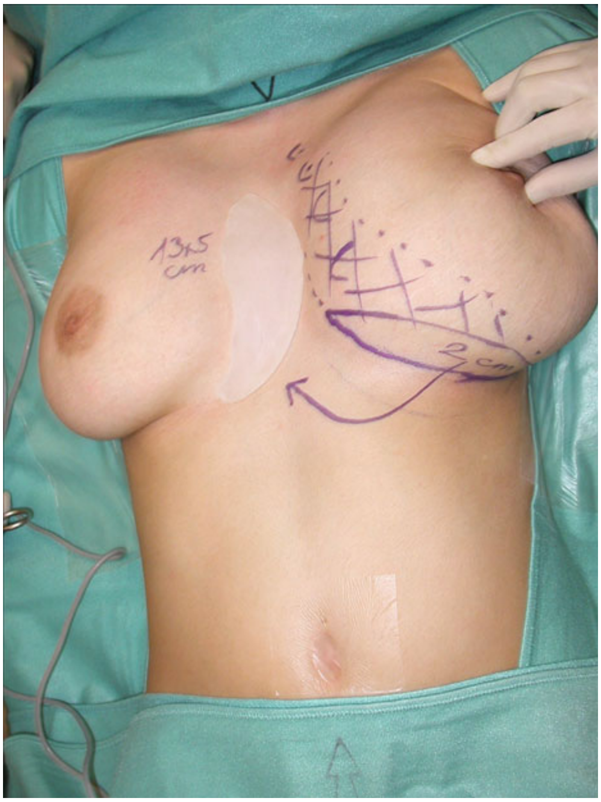

Fig. 2. Intraoperative situs with custom-made (white, semi-transparent, C-shaped) silicone implant already placed on the skin, into the funnel defect. Instead of the silicone implant however, a pedicled flap from the left breast was planned to fill the funnel deformity subcutaneously. The volume of this pedicled breast reduction flap was more than enough to fill up the depression adequately

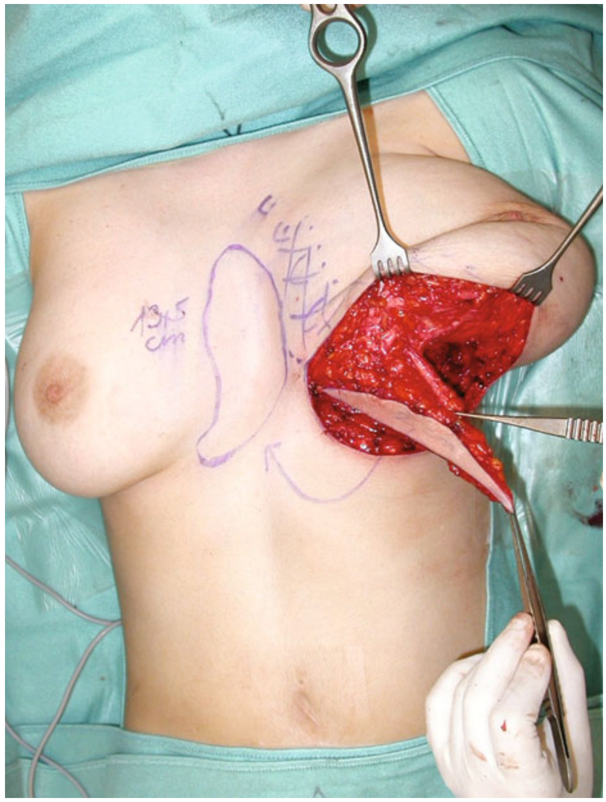

Fig. 3. The pedicled adipo-dermal flap is based on parasternal perforator vessels (indicated by tip of forceps), then de-epithelialized and transposed into a subcutaneous pocket at the presternal funnel. Further surplus tissue of the hyperplastic breast was resected as a discoid from the breast base, resulting in an inconspicuous single scar at the inframammary crease

otherwise surplus tissue and subject of resection during the correction of hypertrophic and ptotic breasts. In this case report such randomly vascularized flaps, based at the lower quadrants of the breasts, were subcutaneously transposed to medial to fill up the pectus excavatum deformity without setting any additional scars. Marshall in 1981 [5] already transposed contralateral breast tissue in oncologic patients, but did this as a two-stage procedure and using a broad medial tissue pedicle. Schoeller in 2001 [11] tried to transpose a contralateral split breast flap for the purpose of breast reconstruction. The relatively large volume flap was based on the perforators of the internal thoracic vessels, but the distal third of the flap had to be discarded due to venous congestion. The pitfall therein seemed to consist in too much tissue bulk that was transposed immediately and based on these perforators, but the anatomical background of that concept gave background to further investigations. Vesely in 2007 [12] and Schmidt in 2010 [10] provided with detailed anatomical studies concerning the internal mammary artery perforators (IMAP) are available, describing its potential use also in central and contralateral chest or breast reconstruction.

In our case described herein, a disk-shaped tissue flap remains vascularized by minor pedicles at the lower medial quadrant and at the basis of the breast tissue cone. This flap is dissected out beginning from lateral and caudal and off the pectoralis major muscle as well as the breast parenchyma. Designed as an island-flap it remains yet nourished (Fig. 3) by several parasternal intercostal perforators medially arising from the internal mammary vessels [12, 13]. This island-flap is de-epithelialized and then transposed to medial. Under circumstances and if the fibroadipose tissue flap presents as a rather stiff tissue bulk especially in adolescents, it can be brought into the

Fig. 4. Very pleasant situation 8 months thereafter without any visible scar, even in an aspect from beneath

funnel depression even as a turn-over flap. It is transposed into the defect and attached there using the same submammary incision by which the subcutaneous pocket extending the funnel cavity is dissected (Fig. 3). The incision at the inframammary crease along which flap harvesting is undertaken will be closed likewise to a reduction mammoplasty with discoid resection [8]. A perfect symmetry however can hardly be obtained (Fig. 4) even with this elegant technique, but two problems simultaneously are settled or minimized herewith. On the one hand hyperplasia of one or two breasts will be reduced and on the other hand the funnel chest deformity may be corrected within a single surgical intervention. The utilization of autologous tissue for filling up a funnel depression means a particular long lasting advantage in contrast to any foreign alloplastic material. It therewith yields a permanent improvement of shape without potential late sequelae after decades, such as capsular contraction, pocket infection or calsifications in the area of the implant capsule. Moreover the skin incision in the area of the submammary crease is designed to allow access both to tissue resection and to flap transposition. Furthermore postoperatively only this single scar remains but usually is completely covered along the inframammary crease by the natural ptosis of the breast. Because only at the cone basis of the breast tissue fat and minimally glandular tissue is harvested, the ability to breastfeed is preserved in any case. However it might happen on the other hand, that during pregnancy these parts of glandular tissue, which are transposed into the funnel, may swell temporarily and postpartally due to lactogenesis, until breastfeeding is ceased. This technique is recommended only in adults after sufficient maturation of the breasts and well-defined submammary crease. It will not be suitable in firm breasts in youth or adolescents without ptosis, furthermore such interventions into growing tissue may result in an unpredictable outcome concerning both the shape of the breast and the funnel deformity. Preoperative mammography or sonography of the breasts is mandatory as a standard routine procedure to exclude any tumor formation prior to use breast tissue for any reconstructive purpose [10, 11].

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6.6.4 Microvascular flaps

Christoph Papp, Wolfgang Michlits

Increasing evidence indicates that funnel chest deformities cause physiologic impairment and limitations, as well as adverse aesthetic and psychological effects [1, 2]. Several techniques have been described to correct this deformity. In the Ravitch procedure, e.g., the deformed costal cartilages are excised, the sternum elevated, and the lower extremity of the sternum supported by perichondrium reefing sutures. Many modifications followed like temporary implantation of Kirschner wires percutaneously, which then had to be removed approximately 12 months after the initial repair [3]. This technique has been modified during decades by some authors using, e.g., a sternal bar (called Adkins strut) for the fixation of the sternum [1, 2, 4, 5]. Nuss described placement of a large curved bar under the deformed chest wall through percutaneous incisions [6]. The bar is kept in place for a prolonged period of time during which the cartilages get reformed in their new position. These two techniques are mainly utilized in children suffering from symptomatic funnel chest to improve cardiovascular and pulmonary function. Ninkovic et al. propagated the free microvascular sternum turnover flap for the remodeling of the thoracal wall in adolescents and adults [7]. Despite these reconstructions that affect the entire thoracic wall are aimed at functional improvement, with all their advantages and disadvantages, some authors described also good aesthetic results using custom-made silicone implants or autologous tissue placed subcutaneously for the correction of asymptomatic funnel chest [8–11]. The indication for the treatment of funnel chest is mainly aesthetic, because up to 95% of the patients do not substantially suffer from cardiovascular or pulmonary symptoms [12]. We therefore prefer the utilization of autologous tissue to correct an aesthetically impairing deformity. At our department we frequently utilize the free groin-flap, the free transverse rectus abdominis musculocutaneous (TRAM)-flap and the free fasciocutaneous infragluteal (FCI)-flap for several other indications of defect reconstruction [13, 14]. Depending on the patient's requirements we select the most suitable flap for each individual patient. Our experience shows that especially in slim patients it is difficult to gain enough autologous tissue volume, e.g., from the abdominal wall or the groin region for an adequate filling of the funnel depression. In those cases and in agreement with the patients we choose to use autologous tissue harvested from the buttock region since this commonly is the only region in the usually leptosomatic funnel chest patients which provides with adequate tissue bulk.

6.6.4.1 Indication setting

Adult patients admitted to our department desiring correction of their congenital stain are first of all informed about the various possibilities in the correction of a funnel chest deformity. Usually many of them refuse implantation of foreign materials, while already informed through different media they prefer correction with autologous tissue. Some patients suffering from a small to medium defect size are selected for the transplantation of a groin-flap, predominately because of its inconspicuous donor site. Others tend to receive an abdominoplasty at the same time of tissue harvesting from the abdominal region, however this situation occurs very seldom as most of the funnel chest patients show leptosomatic habitus. Especially slim patients prefer the buttock region as the only region to gain enough tissue volume for an adequate reconstruction. In our description of the surgical technique we will focus on the FCI-flap because the groin-flap as well as the TRAM-flap are well established and documented flaps that need no further description in detail. Nevertheless both flaps will be mentioned hereby.

Groin-flap

This axial pattern flap, for the first time performed as a pedicled flap by Wood in 1862 is a frequently used free-flap even nowadays. It is based on the superficial circumflex ilium artery, dissection is easy in supine position. The flap is outlined preoperatively in an ellipse design, depending on the size required for the individual defect. The medial apex lies medial to the

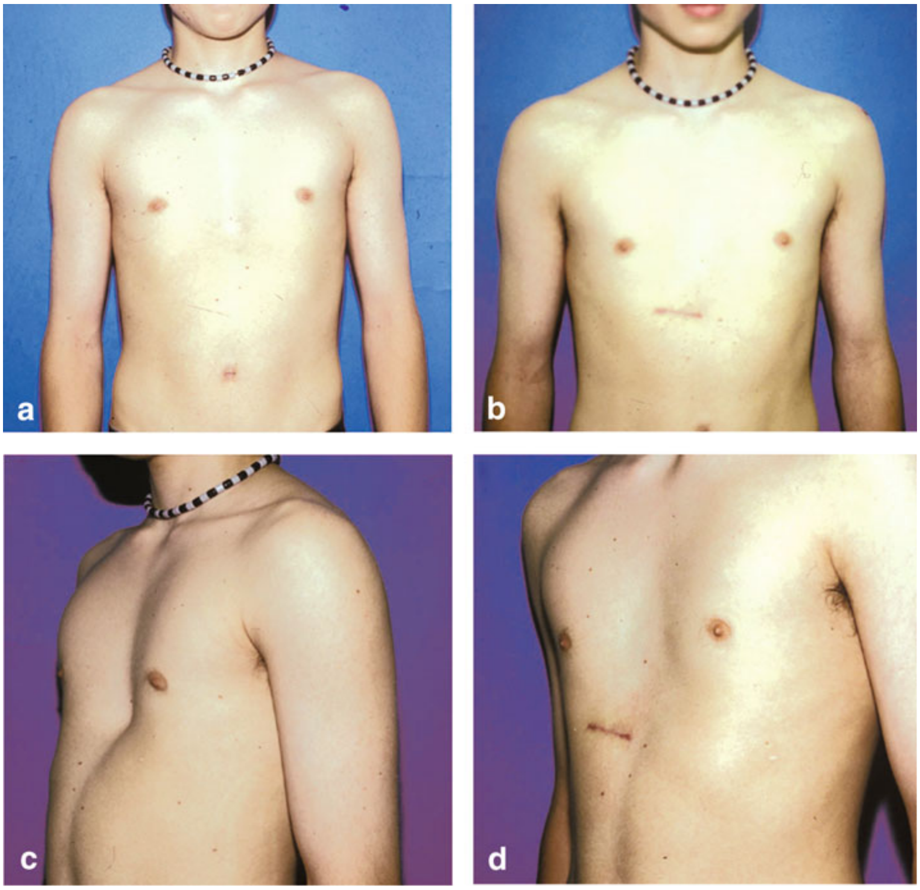


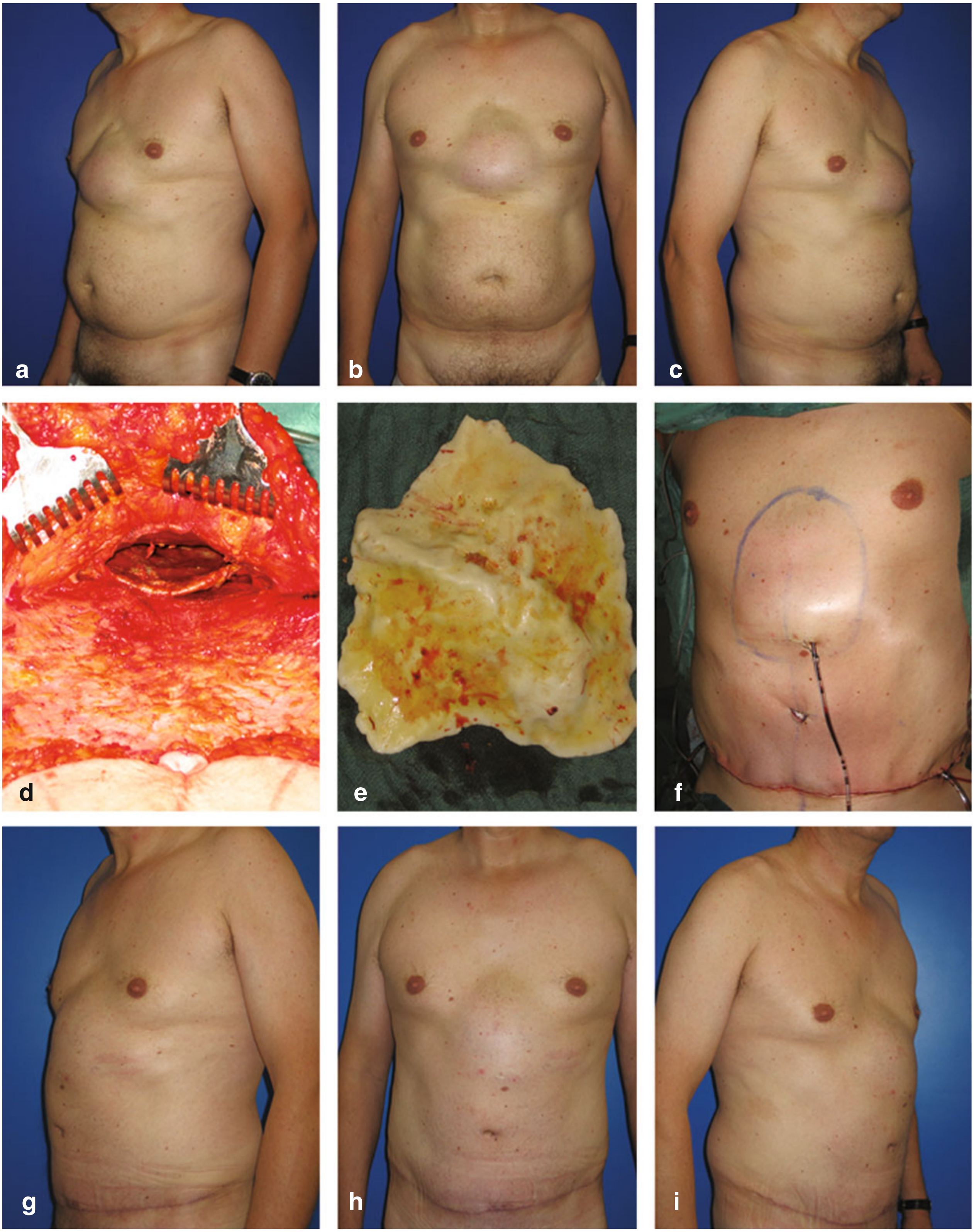
Fig. 1a. An 18-year-old male patient suffering from an asymptomatic PE. **b** Frontal view 3 months after reconstruction. Same patient before **c** and after **d** the reconstruction using the groin-flap in the lateral view

femoral vessels, the lateral apex, depending on the necessary size, may be set as far superior-laterally as the iliac spine. The flap is elevated from lateral to medial. After identification of the vessels they are followed to their origin and separated from the femoral vessels. After deepithelialization not only the flap is anastomosed to the recipient vessels with microvascular techniques and microscope, in most cases to the superior epigastric vessels, but also the internal mammary vessels may serve as recipients (Fig. 1).

TRAM-flap

This flap is a myocutaneous flap containing parts of the rectus abdominis muscle and is based on its blood supply, the inferior epigastric vessels. The TRAM-flap is frequently used as a standard procedure for breast reconstruction after mastectomy. The operation is also performed in supine position. The flap is outlined preoperatively and ultrasound is helpful preoperatively to localize the perforator vessels to the skin. Usually a muscle sparing TRAM-flap is harvested to preserve the rectus abdominis muscle to prevent hernia or bulging of the abdominal wall. For the reconstruction of a funnel chest deformity we never choose a deep inferior epigastric perforator (DIEP)-flap. Although we frequently use this flap in breast reconstruction, in a solely aesthetic indication risk factors should be minimized. It is safer to harvest and transplant a muscle sparing TRAM-flap than a DIEP-flap solely based on tiny perforator vessels.

Fig. 2. A 51-year-old man suffering from a massive capsular formation and infection after reconstruction of a PE with custom-made implants 14 years ago. **a–c** Showing the deformity before the resection of the custom-made implant and the capsular formation. **d** The capsular formation with the implant in situ. **e** Custom-made implant after resection. **f** Patient after reconstruction with the TRAM at the end of operation. **g–i** Outcome 6 months after operation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After flap elevation also this flap is deepithelialized and anastomosed to the recipient vessels at the thorax. Transplantation of too much volume in some cases required flap shaping and/or liposuction about 2 months after the first surgical intervention. Figure 2 shows a patient suffering from a massive praesternal capsular formation and seroma after subcutaneous implantation of a custom-made silicone block 14 years ago. The consecutive puncture of the seroma led to an infection making an operative intervention necessary. The resulting cavity after explantation of the silicone block and the calcified capsule was filled up with a free TRAM-flap.

Free fasciocutaneous infragluteal-flap

The FCI-flap is a fasciocutaneous-flap based on an end-artery, the cutaneous branch of the descending branch of the inferior gluteal artery [15, 16]. The size of the skin island ranges from 10 × 5 cm up to 18 × 10 cm, e.g., in patients receiving a simultaneous correction of breast asymmetry (Fig. 4a, c, e). Ultrasound of the recipient and donor vessels is performed preoperatively in all cases to identify the anatomical situation and mark the cutaneous branch of the descending branch of the inferior gluteal artery preoperatively [15]. The patient is initially placed in the prone position likewise to a buttock lift. The donor area of the subcutaneous tissue harvest will be extended cranially, laterally and caudally from the marked donor skin in the infragluteal crease (Fig. 3a). The fat pad and bursa above the ischial tuberosity is preserved and further harvesting of the flap proceeds from lateral to medial exposing the gluteal musculature. The flap vessels accompany the posterior femoral cutaneous nerve that extends into the subcutaneous fat pad and further caudally along the ischiocrural musculature. The flap vessels pass to the middle third of the gluteus maximus muscle and curse from the edge of the muscle [16]. Separation of the posterior femoral cutaneous nerve from the flap vessels can be challenging, so that dissection should proceed with loupe magnification to avoid any injury to the nerve and/or the vascular pedicle (Fig. 3b, c). After successful preparation of both structures the entire flap can be raised (Fig. 3c). To gain sufficiently long pedicle length the gluteus maximus muscle can be elevated for further vessel dissection. The vascular pedicle can be followed up to the infrapiriformis foramen by ligation of several perforators arising from the pedicle. In most cases there is a more prominent branch of the inferior gluteal artery to the gluteus maximus that originates close to the infrapiriformis foramen. Ligating this branch allows 2–3 cm of pedicle length to be gained. Thereafter, the flap's artery and vein are separated and cut off at the infrapiriformis foramen. After careful, in order to preserve to cutaneous nerves, wound closure the patient is turned to the supine position, and the recipient site at the funnel chest is prepared.

First a 5-cm skin incision is made in the inframammary crease beginning about 3–4 cm laterally from the median line. Then a subcutaneous pocket is dissected carefully in the area of the funnel depression. Thereafter, the superior epigastric vessels or the internal mammary vessels are prepared for anastomosis. Then the flap is deepithelialized, positioned into the defect and microsurgical anastomosis is accomplished. In all our female patients we were definitively successful to improve the appearance of the breast (Fig. 4). Finally, after wound closure loose dressings are applied to avoid any tension or pressure to the flap, which might swell by edema formation. That is why the pocket should provide with ample space in order to prevent self-strangulation.

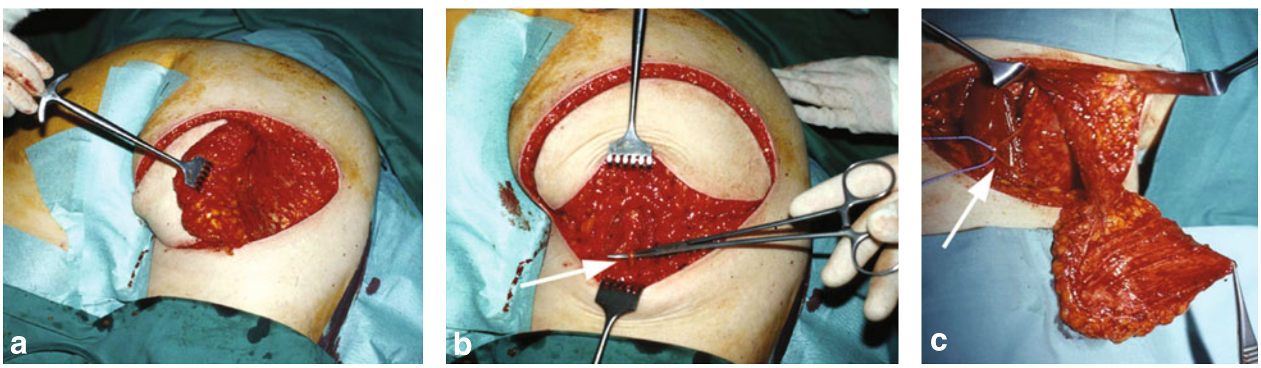

Fig. 3. Intraoperative image of a 26-year-old woman showing the preparation of the FCI-flap in the prone position. **a** Flap preparation is always started laterally. **b** Thereafter, preparation is preceded medio-caudally by identifying the border of the gluteus maximus muscle. **c** Image showing the FCI-flap before the detachment of the pedicle. Arrow is pointing to the posterior femoral cutaneous nerve

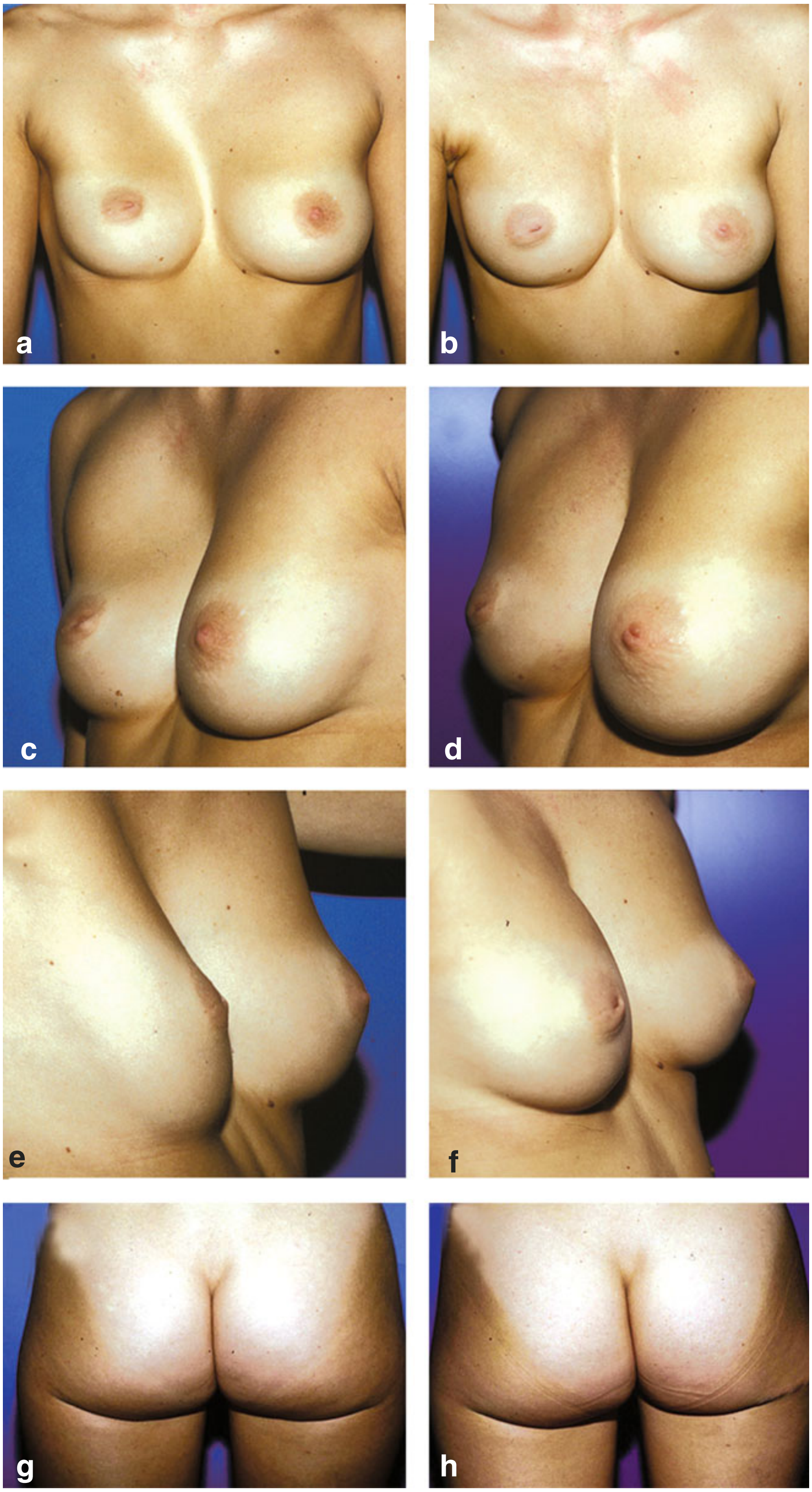

Fig. 4. Image shows the preoperative appearance of a 26-year-old woman suffering from a PE (**a**, **c**, **e**, and **g**). **b**, **d**, **f**, and **h** Images show the outcome 23 months after the surgical correction using the free FCI-flap. Note the rotation and deformity of the right breast preoperatively that is corrected completely. **h** Inconspicuous scar at the donor site

6.6.4.2 Results

A successful reconstruction of asymptomatic funnel chest deformity can be performed using autologous free-flaps from the groin, the abdominal, the buttock or the back region [17]. Operating time in our institution ranges from 210 to 320 min, in our series of funnel chest reconstruction we had no flap loss. None of the patients developed neuroma or seroma at the donor site. Infection or dehiscence of the wounds also was not observed. One patient, after harvesting of a FCI-flap, suffered from a temporary sensory discomfort at the donor site during the first two postoperative weeks that resolved completely within 6 weeks.

Patients were discharged about 7–10 days postoperatively and regularly examined in our outpatient clinic. The flaps that appeared bulky immediately after the inset, probably due to edema, retracted to the desired volume after several weeks. Nevertheless, in singular cases it was necessary to perform flap shaping and/or liposuction of the flap to improve the aesthetic appearance and reduce tissue bulk. These corrective operations were carried out about 3 months after the first operation. Finally all patients were fully satisfied with the result and would undergo the reconstruction with autologous tissue again (Figs. 1, 2, and 4).

6.6.4.3 Discussion

Congenital chest wall deformities are always challenging for the surgeon in consultation as well as in reconstruction procedures. The majority of indications for surgery in the funnel chest deformity are aesthetic and psychological reasons [1, 8]. Several techniques and among them invasive procedures, requiring fracturing and remodeling of the chest wall skeleton are potentially associated with high morbidity and high rate of complications [1–7]. These invasive procedures should be probably reserved for children with severe deformities and accompanying impairment of cardiac and pulmonary function, but still pliable bone and cartilages.

Various techniques have been described to treat patients suffering from asymptomatic funnel chest (approximately 95% of cases). Some authors described excellent outcomes using silicone implants [8, 9, 12]. Nevertheless, some patients refuse the use of foreign materials and prefer the reconstruction with autologous tissue. Therefore, plastic surgeons should offer their patients a few possibilities to find the perfect flap/tissue for the individual patient.

The FCI-flap is ideally suitable for the treatment of asymptomatic funnel chest deformity, the main advantages of this flap consist in the inconspicuous donor site scar situated in the buttock crease. A further advantage is its long vascular pedicle (up to 18 cm) that allows a wide range of motion during inset. Especially in slim patients the FCI-flap is an excellent flap because of the possibility to raise a large skin island together with the surrounding subcutaneous fat tissue. Despite taking a large amount of tissue only a mild asymmetry at the buttocks is visible during the early postoperative period that recovers over time. Our long-term experience with this flap proved that postoperative efforts should focus on meticulous hemostasis and the postoperative application of compression garments to the donor site to avoid seroma formation. Of further importance is physiotherapy to mobilize and stretch the donor site in order to regain pliability of the surrounding tissue.

Moreover, these patients should preoperatively be informed about the sitting discomfort in the early postoperative period.

Finally, in our opinion the reconstruction of the funnel chest deformity using autologous tissue offers an excellent alternative to established more or less invasive techniques with or without foreign material, however in selected patients.

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6.6.5 Free microvascular sternum turnover flap

Anton H. Schwabegger, Milomir Ninkovic

Using the so-called sternum turn-over for the correction of extensive funnel chest deformities in earlier decades, the sternum and the suspending rib cartilages were taken out of the anterior thoracic wall en bloc or otherwise lifted but pedicled at the dorsal periosteum of the sternum, were turned around 180°, and then reimplanted as a free or pedicled autologous graft [9, 10, 28]. Bone resorption in these turned over but non-vascularized tissue grafts [27, 28] leads to inevitable recurrence and therefore were major drawbacks of these procedures. Loss of elasticity of the thoracic respiratory function, caused by fibrosis of cartilage remnants, infection, or dislocation of the implants and thus unimproved physical condition [5, 7, 26] or undesired aesthetical appearance resulted. This was especially problematic at the age beyond childhood, when revascularization and nourishment through embedding tissue were not sufficient any more for viable reintegration of the turned-over tissue.

On the basis of these disadvantageous results, some authors turned over this skeletal tissue block 180° as well, but not only based on the sternum periosteum, but also particularly pedicled at both internal mammary (thoracic) vessels [8–10, 25, 26]. The cross-over of both vascular pedicles, caused by such a turn-over procedure, on the one hand was advantageous for the genuine blood circulation within the graft; however, on the other hand it caused relative shortening of the vessels along the median centerline on the basis of twisting and thus shifting of the sternum and adjacent rib portions cranially, not beneficial for the purpose of thoracoplasty. Until now this method is described solely in single case reports, and long-term observations are lacking so far.

However, and on the basis of further developments in microsurgical techniques, for this type of thoracoplasty nowadays a method of microvascular turn-over of the sternum and adjacent ribs is available [7, 13, 26]. The revascularization of the 180° turned around osteo-chondro-muscular composite graft logically leads to much improved viability of such tissue than it would do without own perfusion. Within small children, this kind of sternum turn-over without revascularization may lead to “graft take” of still very thin skeletal structures yet, but in contrast to that in the adult such a “graft take” is unfeasible in any case. Based on the size and large volume of tissue block such a procedure inevitably leads to necrosis with ensuing absorption, under circumstances even to hazardous infection and furthermore to inevitable recidivation of the funnel deformity.

In addition to these above-mentioned techniques several others are available for the remodeling correction of pectus excavatum, which is in the case of any necessity for expansion of the thoracic cavity in order to improve cardio-pulmonal capacity [1, 3]. The most popular and very established method relies on the MIRPE technique, previously developed by Donald Nuss [14], with implantation of a specially formed metal strut using a minimally invasive access under videoendoscopic control (VATS, Chapters 5.4 and 6.3), but without the necessity of osteochondotomies.

In many cases with rigid skeletal thoracic structures, a method further developed from the method of Nuss and out of earlier methods may also be applied in combination with a semi-open access (MOVARPE, Chapter 6.1 until 6.4).

Above all in adults, the Ravitch method [16] that is without metal strut suspension of the remodeled anterior thoracic wall tissue may lead to relapse in up to 36% [2, 14]. Also established alternative methods [1, 17–19] can lead to relapse, if the supportive metal struts do not remain sufficiently longer in situ or the adolescent or adult thorax has matured to an already too rigid structure that withstands simple internal strain.

The osteochondroplasty or the sternal support with metal struts, combined with a minor open access (MOVARPE, Chapter 6.4), that is with multiple osteotomies and chondrotomies to relief a rigidly formed funnel, may cause extensive fibrosis and scarring in the area of the remodeling. Such fibrosis, if the operation was carried out already at an early age, on the one hand can lead to growth interferences and therewith to an unpleasant deformity of the anterior thoracic wall. As a worst result such scarring may even inhibit further growth of the anterior thoracic wall, but with the thoracic spinal column still growing leading to countervailing kyphosis of the back [4]. On the other hand such extensive fibrosis and scarring can frustrate the attempt of success of the expansion of the thoracic cavity also in adults. Such auto-reparative processes diminish any elasticity within the area of remodeling, so that the therapeutic effect of an improvement of cardiopulmonal function will be impeded therewith [4].

In rare cases with nickel allergy, several different methods containing the procedure of metal strut implantation for temporary suspension of the anterior thoracic wall are not applicable. The manufacturers offered preformation of unbendable nickel-free titanium bars, but they were not practicable especially in adults due to the lack of bending properties, which particularly led to hazardous maneuvers during their removal. Especially in such selected cases the microvascular turn-over technique presents itself as an alternative to circumvent these problems and will be described now in the following.

Ishikawa in 1988 and Chen in 1999 already performed the microvascular sternum turn-over in several cases; Ninkovic in 2002 also presented a small series of selected patients treated with that complex procedure [8, 13, 26]. Through this turn-over of 180°, the existing depression of the sternum is directly converted into an elevation indeed. That artificially emerging keel chest deformity at this composite graft has to be now flattened by multiple chondrotomies, so that an aesthetically appealing result is formed. This procedure moreover yet expands the thoracic cavity and still maintains the elasticity between rib cartilages and sternum, the sternocostal joints, which are synchondroses and diarthroses (Chapter 2.1). Attributable to that microvascular revascularization of the turned-over part of the skeletal thoracic wall, necrosis will be avoided and therewith effects of extensive absorption or fibrosis are absent, so that elasticity necessary for the respiration movements remains preserved. During that surgery parasternal rib cartilages are cut but not resected and the required sternum osteotomy subsequently is fixed in the sense of an osteosynthesis with absorbable material [23]. This kind of osteosynthesis, applied with diligent technique, should lead to physiological bone healing with only slight callus formation. Additionally securing sutures at the rib transsections support the chest stability.

6.6.5.1 Indication setting for that kind of surgery

Based on the yet existent invasiveness and the long duration of such a microvascular intervention, this kind of remodeling is rather reserved for the adolescent and adult patients. However, this indication is valid only if cardiopulmonal restrictions exist and the patient rejects alternative methods or other distinct reasons exclude the use of a metal strut (e.g. nickel allergy, refusal of metals in the body, or multiple preceding intervention). Indication setting for such an intervention is postponed after a psychological investigation, a 3D-CT-scan as well as a medical investigation of circulation and pulmonary functions (Chapter 3.5). Only then and also after careful and extensively conducted informed consent with the patient, this intervention is carried out on an interdisciplinary basis and jointly with a thoracic surgeon. Prerequisites are however abundant experiences with the microvascular technique. Above all exact knowledge and experiences with the internal mammary (thoracic) vessels are also required, for example in microvascular breast reconstruction as with free flaps [6, 11, 12, 20–22].

6.6.5.2 Surgical technique

The presternal skin incision can be made like a lazy S or inverted Y (Fig. 1a and b), which alleviates the access and the exposition of the sunken sternum. This shape finally yet leads to a swung but optically short scar, which occupies only a small area at the anterior thoracic wall. Both pectoralis major muscles are elevated from the sternum and the rib cartilages including small margins of sternal periosteum for the purpose of reinsertion after completion of the skeletal remodeling process. In appropriate manner also the insertions of the rectus abdominis muscles and the linea alba are meticulously elevated from the xiphoid and the inferior rib edges. Subperichondral resection of 1–2 cm of rib cartilages along the depression region now follows until the area where the sternum declines backwards. Exactly here and after relief of perichondrium as well as intercostal muscles the internal thoracic vessels are carefully dissected free and prepared for microvascular anastomoses. However, the backward angulation of the costal cartilages at the sternal margin allows only limited exposure and therefore complicates the microvascular anastomoses [15]. That is why experience in the microsurgical techniques is absolutely mandatory, because also the respiration movement lets vessel suturing under microscopic view become a severe endeavor.

Especially in these patients requiring such sophisticated kind of remodeling, the access to the vessels is usually located at the level of the second or third rib. From cranial to caudal, the rib cartilages and the intercostal muscles in the area of the funnel depression will be transsected. The thoracic surgeon now carefully mobilizes the intrathoracal mediastinal structures and organs from the back of the sternum and rib cartilages. If feasible, the parietal pleura is dissected carefully from the osteochondral thoracic wall block, without opening the pleural cavity, which means an advantageous epipleural dissection. With regard to avoidance of pleural

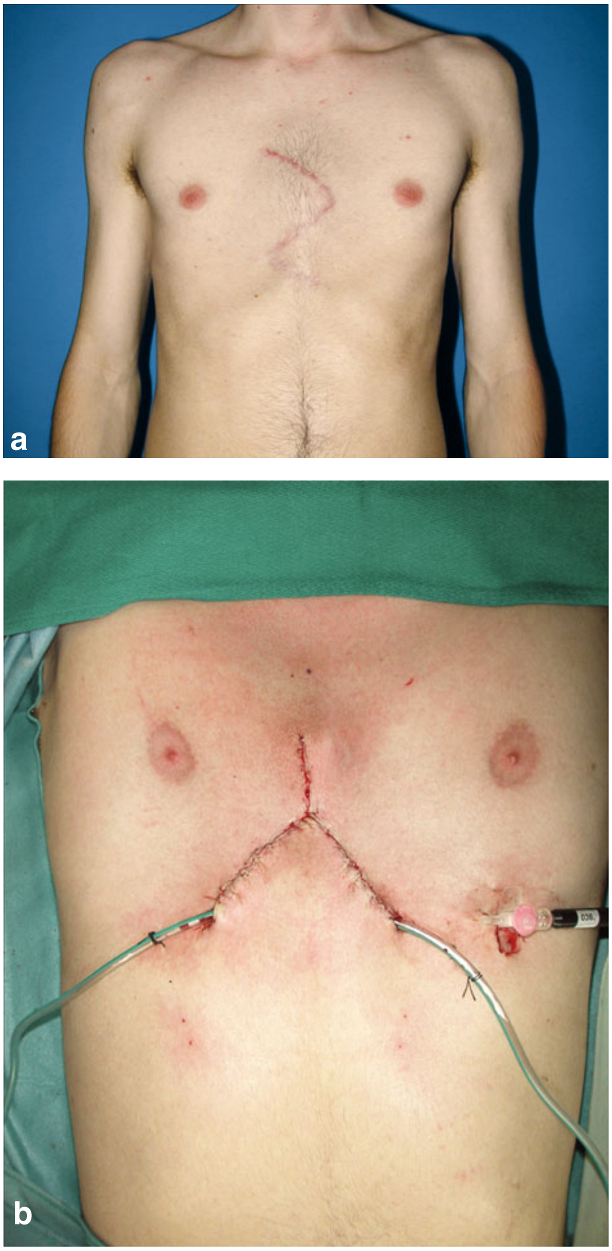

Fig. 1a. Alternatively used curvilinear incision for a better survey of the area of remodeling and the important zone of microvascular anastomoses at the right side at the level of the second intercostal space in this case. **b** An inverted "Y" incision may suffice, if the access for microvascular anastomoses due to advantageous anatomical situation of the deformity may be placed more caudally. Notice pO_2 probe at the left side

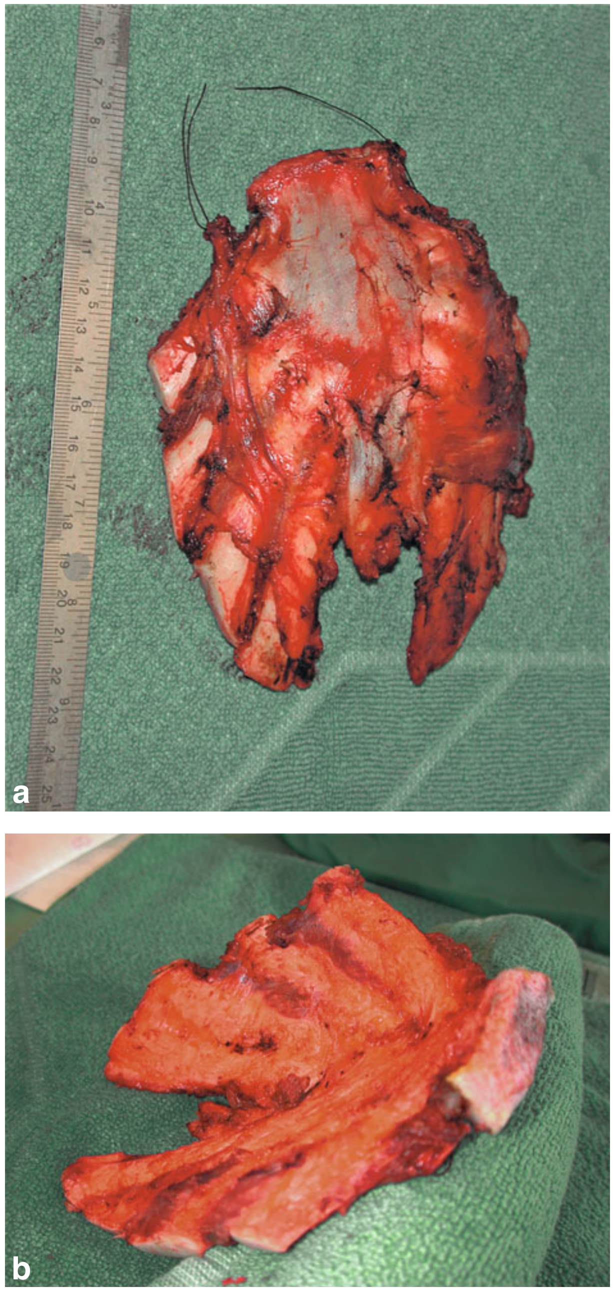

Fig. 2a. Harvested composite flap, containing the inferior thirds of the sternum, the adjacent ribs (four to seven bilaterally), and the internal thoracic vessels as the microvascular pedicle prepared for anastomoses, marked with a suture. View from dorsal. **b** Similar specimen, view from an aspect cranial and the funnel faced anteriorly

leakage and ensuing danger of pneumothorax and/or hematothorax, the integrity of the parietal pleura should be preserved unconditionally. After mobilization of the sternum-rib cartilage-block now follows the preparation of the internal thoracic vessels, so that on one side the recipient vessels and on the other side the donor vessels of the so-called ostoechondral flap are dissected with some excess of length. The surplus of vessel length herewith is necessary to provide with sufficient length of tissue for manipulation after the turn-over procedure (Fig. 2a and b).

After the vessel preparation, the sternum will be cut horizontally with an oscillating saw at the level of the corresponding intercostal area. The circumcised block containing the sternum and its adjacent rib cartilages is now elevated as a free flap (Fig. 2a and b) and is turned around 180°, with the inner surface now turned outwards. In order to avoid the now artificially created keel deformity resulting from that turn-over procedure, single chondrosternal synchondroses and diarthroses can be resected, if necessary for appropriate remodeling. However, some of those spurious joints must be preserved in order to maintain sufficient elasticity for respiration movements. To maintain adequate elasticity, parasternal chondrotomies should be carried out primarily and prior to any flexible joint resection. Especially during these manipulations, the internal thoracic vessels must be protected with extreme caution, since only these vessels are capable of maintaining viability of the turned-over graft. To secure now the molded physiological flat contour of the turned-over skeletal tissue flap, absorbable osteosynthetisis plates can be mounted at the docking areas of the transsections of the rib cartilages (Bionx^{TM} Implants, Tampere, Finland).

At the site of sternum osteotomy absorbable synthetic material is also used in order to obtain a solid osteosynthesis (Figs. 3a and 4). The corresponding rib cartilages are fixed with PDS mattress sutures thereafter in order to obtain additional parasternal lateral stabilization. Now the internal thoracic vessels of the turned over graft are anastomosed to the recipient vessels in microsurgical technique and with the aid of an operation microscope. The setting of anastomoses is possible only at the side where surplus length of the vessels is prepared. Because this microvascular turn-over is a matter of a buried flap, a pO_2-probe (Licox-GSM, Kiel, Germany) or similar device has to be implanted in order to enable continuous measurement of the vascularization and thus viability during the first postoperative week (Fig. 1b). In the rare case of impaired perfusion, the potential complication of tissue loss may

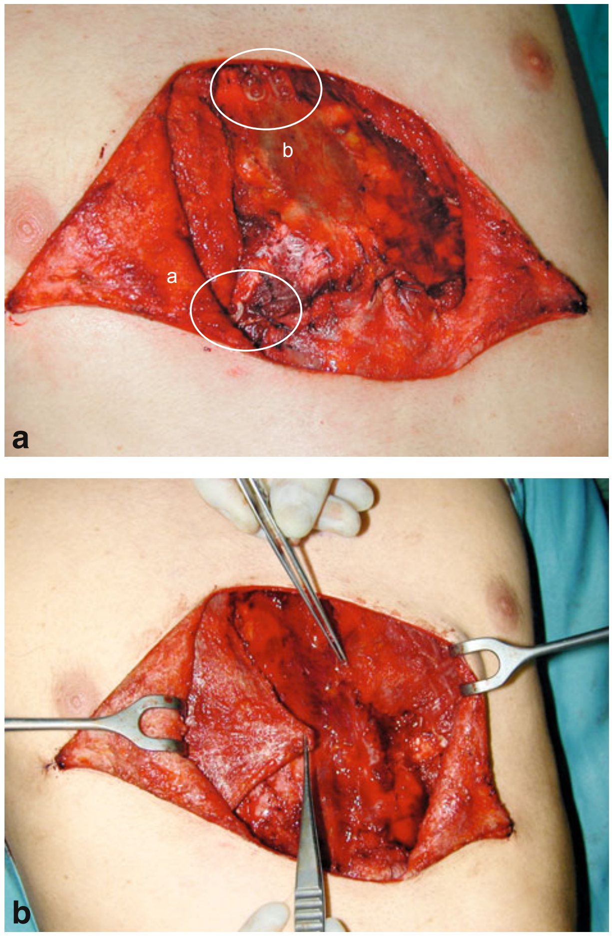

Fig. 3a. The turned-over composite flap already turned over and revascularized by microanastomoses to the internal thoracic vessels, prior to muscle flap reposition and wound closure. Circles show bio-absorbable osteosynthetic material at the rib cartilages (*a*) and the sternum (*b*). **b** The previously elevated pectoralis muscle flaps are anatomically repositioned and tightly fixed to the posterior sternum periosteum

be recognized timely and appropriate microvascular revision can be performed [29].

Subsequently the careful anatomical repositioning of the elevated pectoralis (Fig. 3b) and rectus abdominis muscles is then performed at the sternum to the periosteum as well as to the perichondrium of the ribs. Now, due to the turn-over procedure, the internal thoracic vessels are ventrally exposed, particular caution has to be paid during the anatomical refixation of muscle with seaming sutures along the course of the vessels. Herein compressing seams must be avoided in the environment of the vessels in any case. For the case of any pleural lesion during the flap harvest, the use of a chest tube is recommended. The implantation of a chest tube is

Fig. 4. Solid osteosynthesis of the sternum at the level of the second intercostal space, 6 years after the turn-over procedure

meaningful anyway in order to divert seroma formation, pleural effusion, or even hemorrhage, which is to be expected in such invasive interventions with moderately high incidence and must not be neglected.

6.6.5.3 Discussion

The non-vascularized sternum turn-over described by Wada [28] in any case is not suitable in the remodeling of an adult thorax because the very large tissue block cannot sufficiently become revascularized through the covering soft tissue. The unavoidable consequence would mainly be necrosis and major tissue absorption, in further consequence also potential infection of the large skeletal tissue block and therefrom-resulting further complications. Such an expanded dissection and devascularization of tissue with ensuing absorption processes then will lead to an expanded fibrotic scar pad, which definitely restricts the pliability of the anterior thoracic wall [4]. Based on such pathologic processes to be expected and seen in a long-term aspect, a non-vascularized sternum turn-over appears to be a clear contraindication for tall or athletic adolescents and adults. The alternative and further developed method of the microvascular revascularization of turned-over tissue leads to a rather physiological healing process with much less development of expanded scars. Furthermore, the sternocostal junctions remain viable thus maintain their function, because absorption processes hardly occur. However, and so far no larger series of the microvascular sternum turn-over technique excepts minor clinical series [7, 13, 26] may be found in the available literature. This is possibly true because well-chosen expertise and narrow interdisciplinary cooperation with thoracic surgeons must meet and meanwhile alternative, less invasive methods are established and were further developed.

Through the development of the minimally invasive (MIRPE-) method according to Nuss [14], many patients sooner and rather decide to undergo this minimally invasive intervention. Above all if the funnel is present flat and less marked, the MIRPE method appears the most advantageous. The microvascular sternum turnover however is rather subject to complications based on its invasiveness why as an early complication pneumothorax, hematothorax, infections, or the danger of the complete flap loss may appear. Especially because of the consequences of a possible flap loss, extensive microsurgical experience is a prerequisite in order to reduce herein the technically involved possibilities of any complication to the most minimal. Possible and until now only in individual cases observed long-term complications exist [24], namely instability of the osteosynthesis and the chondrocostal sites of coaptation. Thereby on the one hand callus formation at the posterior surface of the sternum (Fig. 5) may develop and on the other hand through dislocation of cut rib ends

Fig. 5. Persistent callus formation at the posterior level of sternum osteosynthesis, prone to cause adverse hemodynamic effects on the greater vessels and heart

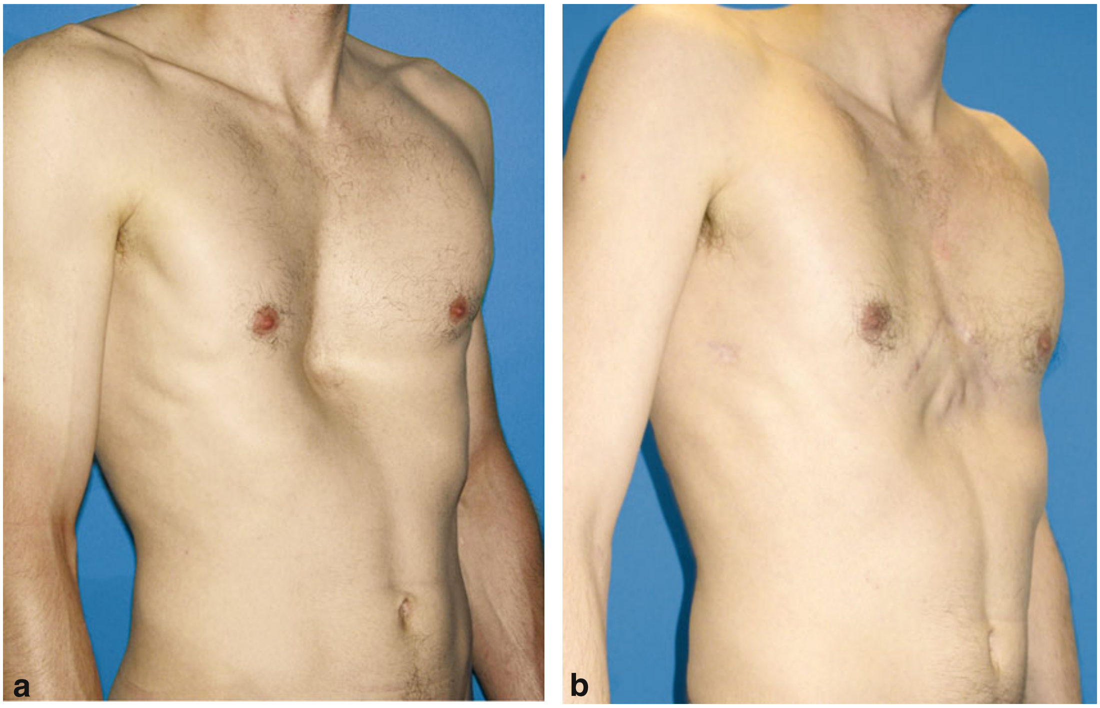

Fig. 6a. 30-Year-old male with moderate cardiopulmonal impairment due to the funnel chest deformity, prior to surgery. **b** Situation 26 months thereafter with stable and rigid anterior thoracic wall and improved aesthetic appearance

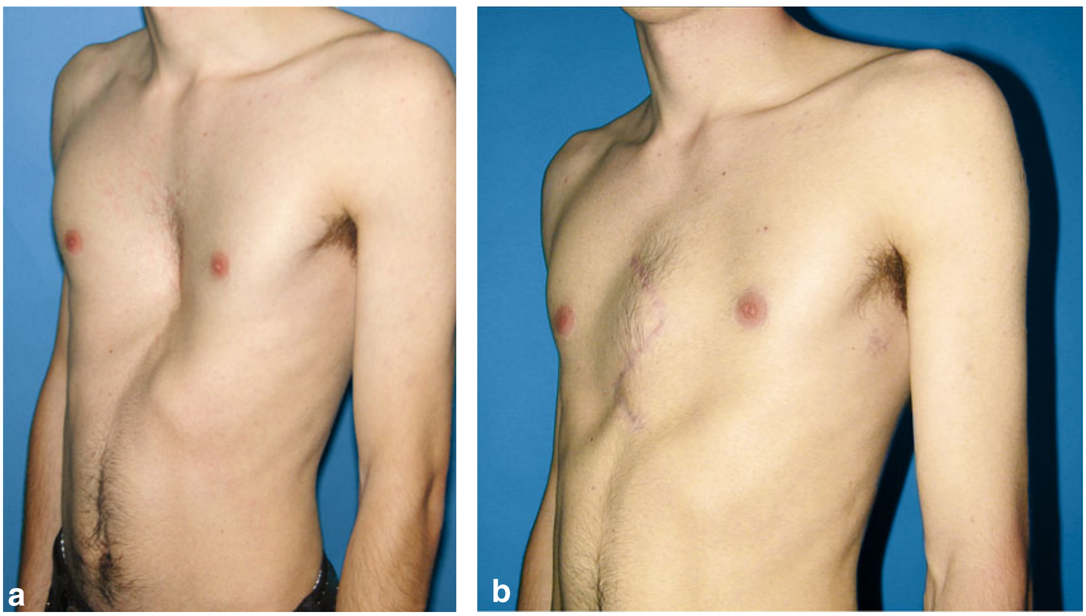

Fig. 7a. 22-Year-old male with sternal depression causing moderate cardiopulmonal restriction. **b** Situation 6 months postoperatively, with inadvertent overcorrection resulting in a slight sternum hump, however, the patient felt very content with that result

compression of the large vessels and the ventricles (Fig. 8b) with potential effects on cardial hemodynamics may follow.
In the few case series described until now [7, 13, 26] the long-term results in regard to aesthetics are described as excellent (Figs. 6a,b and 7a,b). Long-term investigations with regard to alteration or improvement of the cardiopulmonal function or other morphological long-term consequences are not reported up to date. In the own patient series, two patients after a long-lasting period without problems and first after several years complain chronic pains in the surgically involved area of the parasternal end branches of the intercostal nerves. Through the worldwide triumph of the MIRPE technique method, the further development of the microvascular sternum turn-over flap, based on its invasiveness, the required surgical expenditure and the necessity of an extensive microvascular expertise receded into the background. However, indications can still remain, above all in the asymmetrical cases, when the MIRPE or other methods of suspension with or without metal strut will not suffice for the desired shape of remodeling. Especially in unilateral hypoplasia of the thoracic wall, suspension techniques with the pectus bar are not feasible to improve symmetry. They cannot be successful, because the important suspension point for the bar, which should carry the mobilized sternum, is situated too deep at the site of concern. Such an augmentation of a unilateral depression of the anterior thoracic wall appears only feasible through remodeling in the sense of a skeletal tissue exchange from the one to the other side.
The only meaningful and practicable alternative herein exists in the microvascular turn-over because it more or less exchanges the pseudo-surplus of autologous viable tissue from the one side to the hypoplastic other side of the thorax and symmetry assimilation takes place (Fig. 8a and b). Particular care has to be applied in order not to harm the end branches of the intercostal nerves and to avoid the late complications of cumbersome nerve stump neuromas.
For the case that a very steep and deep funnel deformity in adults is present, nowadays the method of the MIRPE combined and modified with a minor central open access offers itself (Chapter 6.4, MOVARPE technique). In contrast to the microvascular sternum turn-over however, two interventions are necessary, on the one hand the suspension of such in parts invasively remodelized thoracic wall by means of an implanted pectus bar and on the other hand its explantation a year or more thereafter. The comparative invasiveness of the MOVARPE technique and the cumulative duration of the morbidity of both interventions vis-à-vis the microvascular turn-over technique appears likewise pains taking for the patient. A comparison of both methods however is not yet possible to date based on the low case numbers of these techniques.

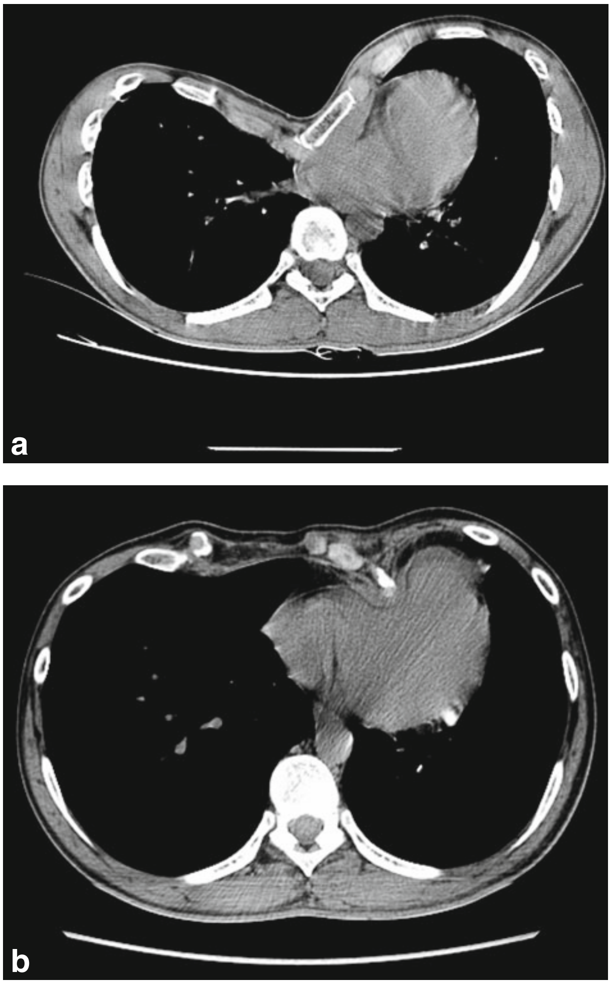

Fig. 8a. Unilateral asymmetric depression with sternum malrotation and reduction of anterio-posterior diameter at the affected side of the thorax, an ideal indication for the turn-over procedure. **b** Same patient as in **a**, 7 years after the sternum turn-over procedure. The cut and refixed ends of the ribs seem to cause an impression at the cardial ventricles with a compressive effect to them, but could not be verified through cardial sonography and functional testing (according to Chapter 3.5). Notice the excellently achieved symmetry of the thorax diameters

Though it appears that still indications remain to apply the microvascular sternum turn-over, in priority the desire of the patient for a single intervention, the refusal of metal implants, or objection to multiple scars, but in particular in an extensively and asymmetrically shaped thoracic wall. It must be considered

Fig. 9. Exemplary case with unilateral and deep depression, massive sternum malrotation and rather not suited for the MIRPE procedure. Alternatively to a suitable MOVARPE procedure, the microvascular sternum turn-over procedure may also serve to improve this situation

indeed in girls and women that herewith a large visible scar stains the zone of the décolleté then. Therefore, the utmost remaining clear indication seems to be the distinct asymmetrical and unilaterally deeply shaped extensive funnel chest deformity in males with proven cardiopulmonal restriction (Figs. 8a and 9). Hereby the previously described experiences are based however on smaller series and further prospective studies with this kind of surgical correction are needed for the consolidation of such an indication setting, however, being well considered and with an informed consent thoroughly elaborated.

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6.7 Combined surgery with associated anomalies or disease

Anton H. Schwabegger

The most frequent and therefore most important monogenic syndromes that go along with pectus deformities are Marfan syndrome, Noonan syndrome, and disruption sequences like Poland and Moebius anomaly as well as genetic associations in exemplarily the Pentalogy of Cantrell (Chapter 2.2).

Pectus excavatum is a hallmark in Marfan syndrome. Pure Marfan or marfanoid features (Fig. 1a–f) are more frequent in the female population and a high percentage, between 50% and 70% of all of them develop mitral valve prolapse [4]. Although stated by Arn in 1989 [1] that Marfan patients operated on pectus excavatum deformities show more relapses, Redlinger in 2009 contradicted this statement on the basis of 246 patients treated so far. Redlinger and co-workers concluded that Marfan and marfanoid patients show a higher complication rate concerning wound healing, which was attributed to connective tissue disorders. There was no difference in clinical outcome of pectus excavatum repair of Marfan or marfanoid versus patients without such disorders in terms of relapse or patient satisfaction. However, usually and probably based on connective tissue disorder the severity of the excavatum deformity in the Marfan and marfanoid collective is higher, requiring more invasive remodeling in the sense of implantation of multiple pectus bars. There is lacking evidence so far, if Marfan and marfanoid patients with a more severe deformity with respect to cardiopulmonal function would benefit more from remodeling surgery than the non-marfan counterparts. However and without evidence, Redlinger and co-workers in their report conclude that the patients felt much better than before surgery. This effect might be based on a psychological effect solely, since pectus

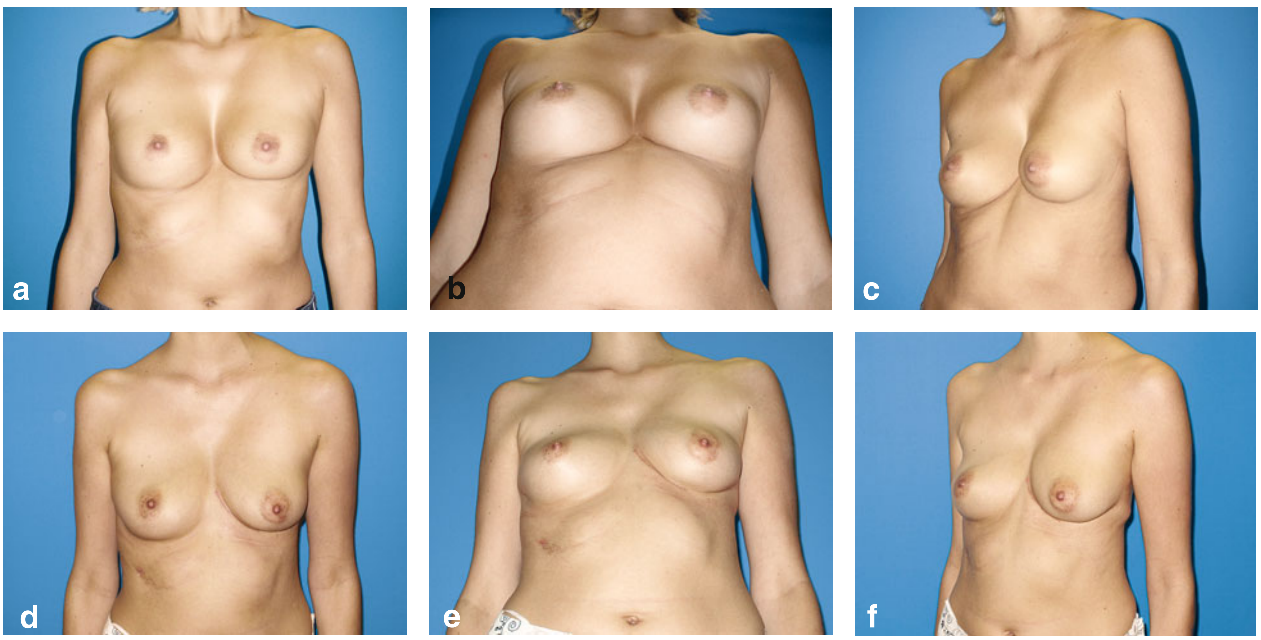

Fig. 1a–c. 26-Year-old tall female with pectus excavatum, marfanoid features, and vertebral scoliosis. **d–f** Same patient, 6 months after implantation of one pectus bar and remodeling via a minor open videoendoscopically assisted repair (MOVARPE)

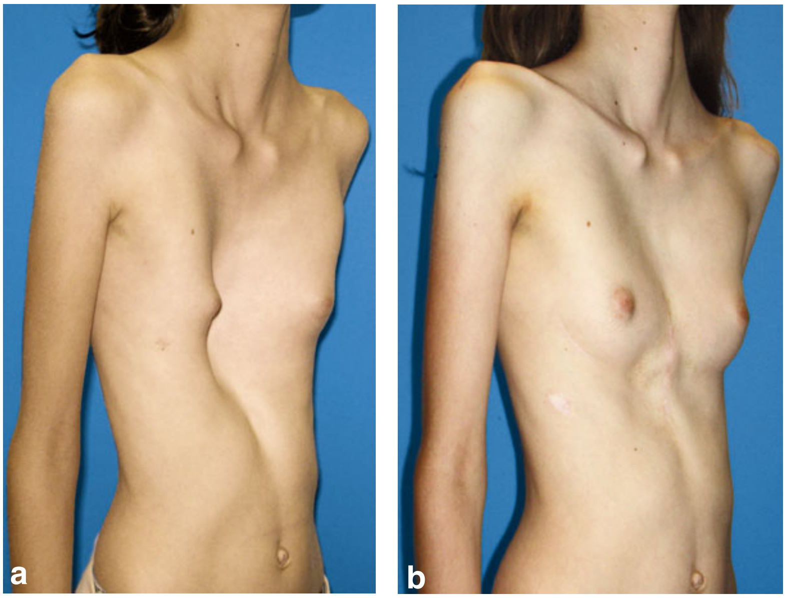

Fig. 2a. 13-Year-old female with severe pectus excavatum deformity and Marfan syndrome. **b** Same patient $1\frac{1}{2}$ years after correction with implantation of one pectus bar and the MOVARPE procedure

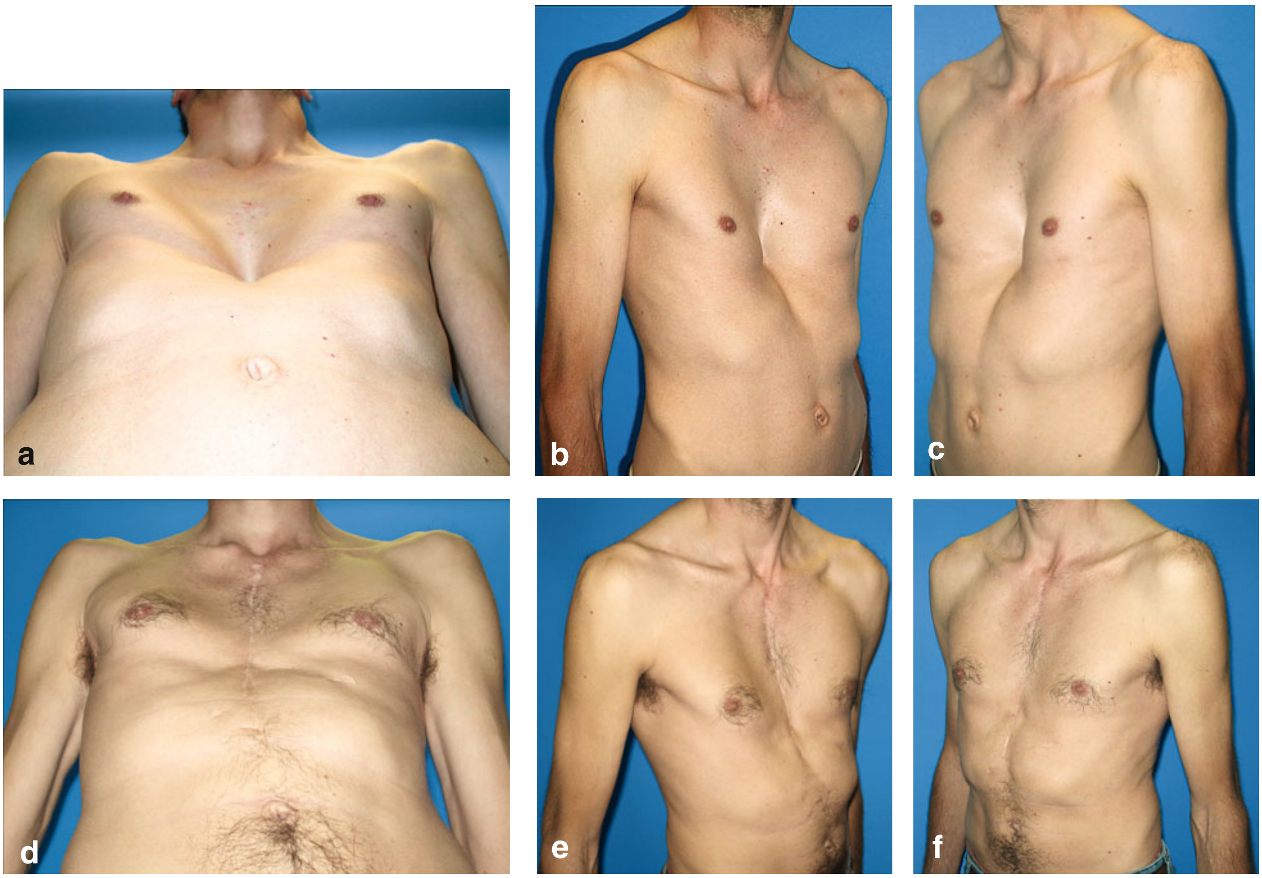

Fig. 3a–c. 38-Year-old male with mitral valve insufficience based on the pectus excavatum deformity. The patient and the thoracic surgeon at the time of cardiac surgery requested simultaneous repair of the deformity to widen the intrathoracic space. **d–f** Same patient 4 years after simultaneous mitral valve and pectus excavatum repair with a modified Ravitch procedure and osteosynthesis with bioabsorbable plates

excavatum deformity in slim and leptosome Marfan or marfanoid patients additionally due to lacking subcutaneous fat tissue presents more dramatically and the results of remodeling procedures therefore seem to be more impressive (Fig. 2a and b) than in non-marfan counterparts.

Occasionally patients with Marfan or marfanoid phenotype present first with the need of cardiac surgery at a higher age. Until then these patients do not bother about their deformity and thus required no remodeling surgery. At the time when cardiac surgery is indicated, exemplarily due to mitral valve insufficiency, the pectus excavatum deformity may be corrected simultaneously. Such a simultaneous approach may even be advantageous for the cardiac surgeon, as a wide exposure of the heart after rib cartilage resection, multiple chondrotomies and sternum osteotomies loosens the severely deformed anterior wall complex, which alleviates the surgical access to the heart [2–6]. A median sternotomy with multiple parasternal chondrectomies at both sides is done. One or multiple horizontal sternotomies are performed to elevate the bony depression for a superior exposure to the usually dislodged heart and intrathoracal organs [5]. After completion of ensuing heart surgery, the remodeling process follows the principles of Ravitch with reefing sutures of the perichondrium tubes. The procedure may be modified in utilizing bioabsorbable plates and screws to refix the osteotomized sternum (Fig. 3a–f). Conventional steel wires, if necessary may secure the fitting. However in the pectus excavatum deformities overabundant length of the rib cartilages and perichondrium tubes allows an almost tension-free closure of the median sternotomy. Such absorbable osteosynthetic material is preferably used because in the case of sudden intrathoracal complications an immediate uncomplicated surgical access is feasible, which would not be the case after utilization of metal plates. Even resuscitation will not be impeded by the malleable biological plates, which on the other hand could become an endeavor in the presence of heavy metal hardware [7]. If a patient must undergo secondary interventions at the heart years later, it might be fraught with technical difficulties due to scar adhesions in an untreated funnel deformity and survey is impeded by the still present sternal depression. The additional remodeling procedure herein requires approximately one additional hour in addition to the time taken for heart surgery. On the other hand, wide exposure through the remodeling access allows a more secure intervention for the cardiac surgeon thus such a simultaneous repair is justified, in addition also with regard to the long-term benefits [5].

The Noonan syndrome phenotype is characterized by normal measurements at birth, short stature later in life, congenital heart defects with most frequently pulmonary valve stenosis and/or cardiomyopathy, broad or webbed neck, pectus arcuatum (Fig. 4a and b), cryptorchidism, coagulation defects, and facial dysmorphisms such as ptosis, wide-spaced eyes, and low-set and posteriorly rotated ears. The surgical correction of such a mixed deformity of the anterior

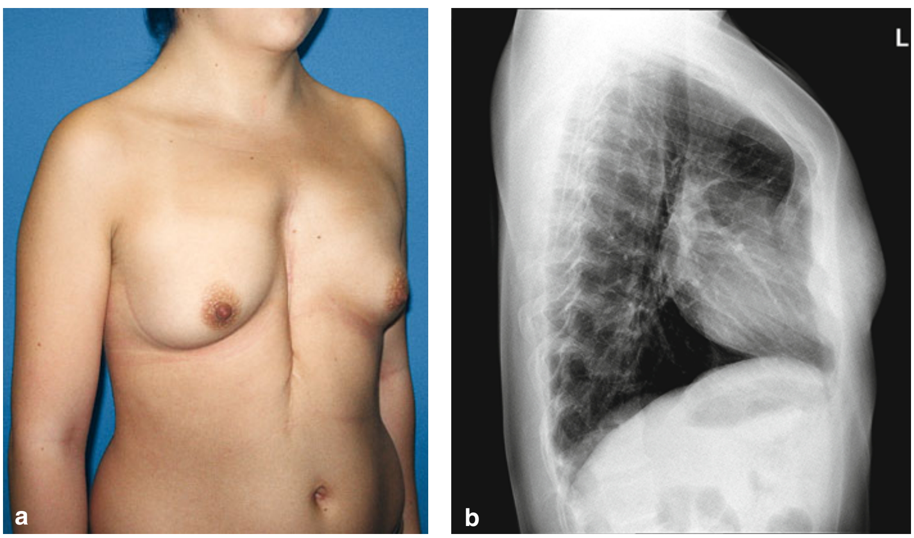

Fig. 4a and b. 21-Year-old female with Noonan syndrome after cardiac surgery in early childhood. Also note phenotypically broad webbed neck and multiply arcuated sternum thoracic wall called pectus arcuatum deformity is described in detail in Chapter 8. Occasionally these patients require plastic surgical treatment for their additional anomalies also.

The extremely rare coincidence of pectus excavatum and gynecomastia may be purely accidental. However the scar that is produced by subcutaneous mastectomy may simultaneously serve as a surgical port to implant a custom-made silicone block via subcutaneous tunnel to fill up the depressed sternal area (Fig. 5). On the other hand and for the case of timely different occurrence of the tumor, the implantation scar may serve for the mastectomy procedure if primarily set in an advantageous area.

More frequently asymmetric deformities at single ribs are present in coincidence with a pectus excavatum deformity (Fig. 6a). If located advantageously and if requested by the patient, the aesthetic correction of isolated rib humps simultaneously with silicone implantation may be managed from the same incision that is primarily placed above the protruding ribs. From the same incision, and after completion of cartilage and rib bone resection, a subcutaneous tunnel is created to implant the silicone block (Fig. 6b and c).

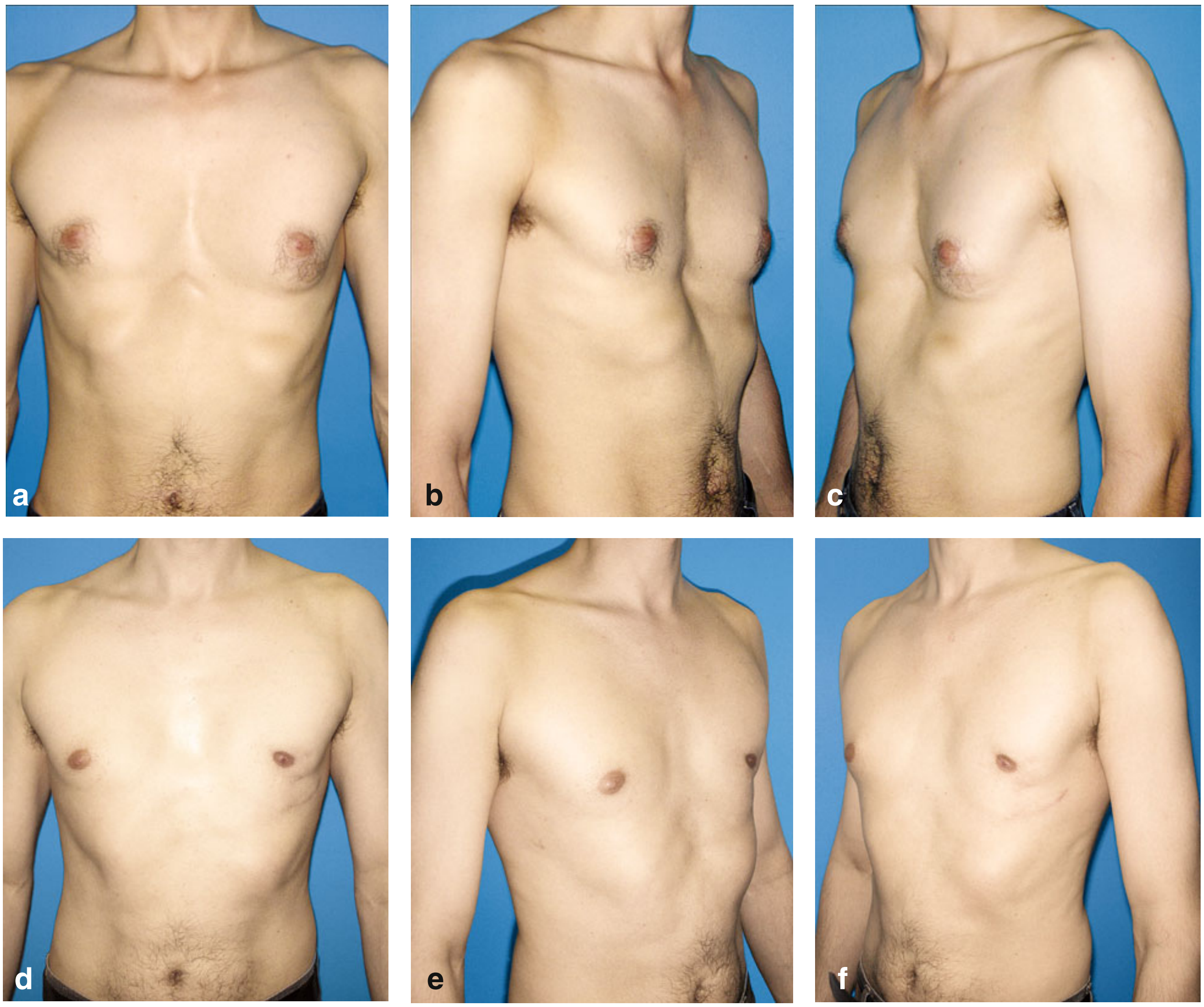

Fig. 5a–c. 25-Year-old male with a unilateral benign gynecomastia at the left side and coincident symmetrical pectus excavatum deformity without cardiopulmonal restriction seeking for aesthetic improvement of both deformities. **e–f** Same patient 7 years after simultaneous mastectomy at the left side, using the same surgical incision for simultaneous silicone implantation at the central presternal region

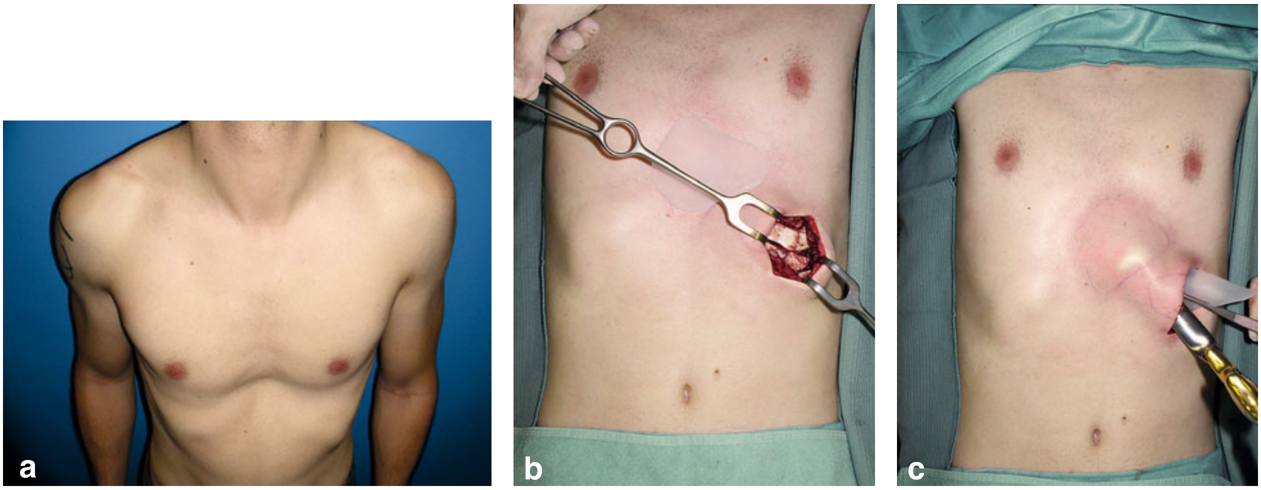

Fig. 6a. 31-Year-old male with minor asymmetrical pectus excavatum and unpleasing rib hump deformity at the left costal arch seeking correction of both deformities. **b** Same patient intraoperatively after completion of subperichondrial rib hump resection. **c** Same patient using the same surgical access for subcutaneous custom-made silicone implantation

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6.8 Vacuum bell procedure according to Eckart Klobe (nonsurgical)

Micha Bahr

6.8.1 Introduction

The conservative therapy of funnel chest deformities has a long tradition. Several of these treatments are described [3, 4], but most of them do not really improve the contour of the thoracic wall.

In 1992, the funnel chest patient Eckart Klobe, desired an improvement of his own deformity, but declined to undergo surgery and developed a special device, which he named vacuum bell, for the therapy of his pectus excavatum deformity. During a period of 2.5 years he was able to elevate his funnel with this self designed vacuum bell. This bell was built with silicone and plaster of paris, bought in a do-it-yourself-store and it was evacuated with a water jet pump (Fig. 1). After 30 months the thoracic wall was elevated to an extent that no funnel was visible any more.

After that success, based on a single own experience, he started to design a professional vacuum bell for a non-surgical funnel chest therapy. Since 2000 he searched for physicians, who wanted to gain experiences with this non-invasive method of deformity correction, but nobody was interested.

The idea to improve funnel chest deformities with negative pressure was not new. In the beginning of the 20th century Professor Hans Spitzy from Vienna and Professor Fritz Lange from Munich already intended to correct funnel chest deformities with so-called vacuum bells. They used a glass bell and could demonstrate that the funnel depression was elevated very quickly during the bell application, but as a drawback the skin was injured because the materials were not suitable for such kind of a therapeutic application [6].

Many patients with funnel chest deformities desire and literally seek for methods to improve the aesthetically annoying stain of their thoracic wall, but they are reluctant to undergo surgery because of the reported pain associated with postoperative recovery and the risk of imperfect results with remaining scars. In many cases of pectus excavatum the degree of pectus deformity does not immediately warrant surgery, however, patients more than ever request some kind of non-surgical treatment.

For these selected patients in 2002 we started to use the vacuum bell according to the inventor Eckart Klobe to improve the funnel chest deformity. With the following years more and more departments in Europe started to use the vacuum bell procedure, so that nowadays it has developed to a common non-surgical therapeutic alternative versus invasive surgery in specialized centers [1, 2, 5].

Fig. 1. The prototype of the vacuum bell

6.8.2 The vacuum bell according to Eckart Klobe

The body of the vacuum bell is made of a silicon ring and a transparent polycarbonate window. At the window an adapter for a simple suction bulb is fixed, designated to evacuate the air from the bell. The bell itself is produced and available in three sizes: “small” for children up to 8 years, “medium” for children up to 12 years, and “large” for children and adolescents elder than 12 years and adults. Since 2 years a vacuum bell for female patients with the shape of an eight is available. This shape provides with the facility to apply this kind of treatment in female patients even during and after full development of their breasts. The

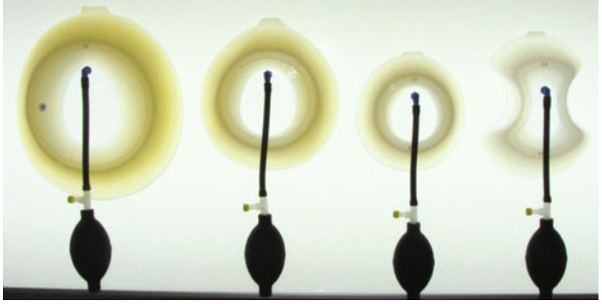

Fig. 2. Variants of the vacuum bell according to Eckart Klobe

vacuum bells of Eckart Klobe are certified, patent-registered, and are liable to the German medical standards (Fig. 2).

The vacuum bell works with two mechanisms. First, there is the negative pressure, which elevates the sternum toward the cavity of the bell. Second, to improve the contour of the deformed ribs, it is essential to apply some kind of pressure at the areas around the funnel or the bell cavity, which is the border of the funnel deformity. This pressure is produced by a silicon ring around the bell cavity, molded to the usually irregular surface of the thoracic wall. Both mechanisms provide with the contour that should be achieved to the thorax.

Before the development of the vacuum bell therapy it was hard to believe that it could be possible to elevate a sunken sternum with vacuum power alone. To prove evidence of this mechanism, CT scans of a thorax with funnel chest deformity were made, first without and then with the application of the vacuum bell. The time period between the situation without bell application and the second CT scan with evacuation took only 2 min. The second CT scan definitely showed that the sternum was elevated and the distance between the vertebral bodies and the sternum was elongated about 30% (Figs. 3 and 4).

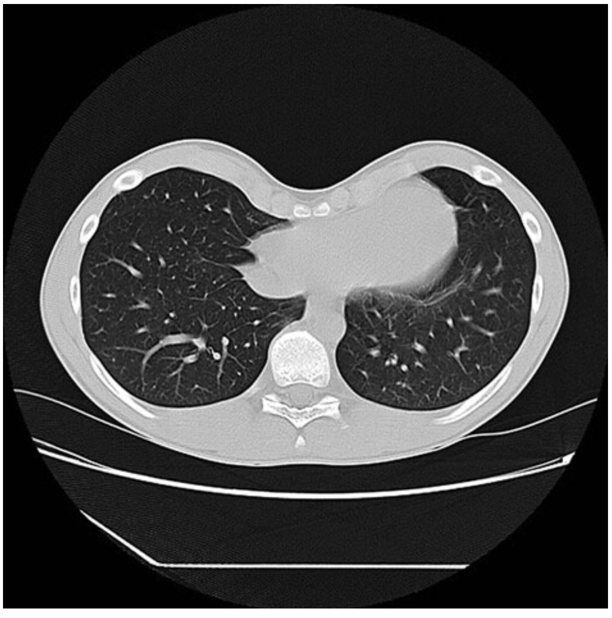

Fig. 3. CT scan of a funnel chest patient without vacuum bell

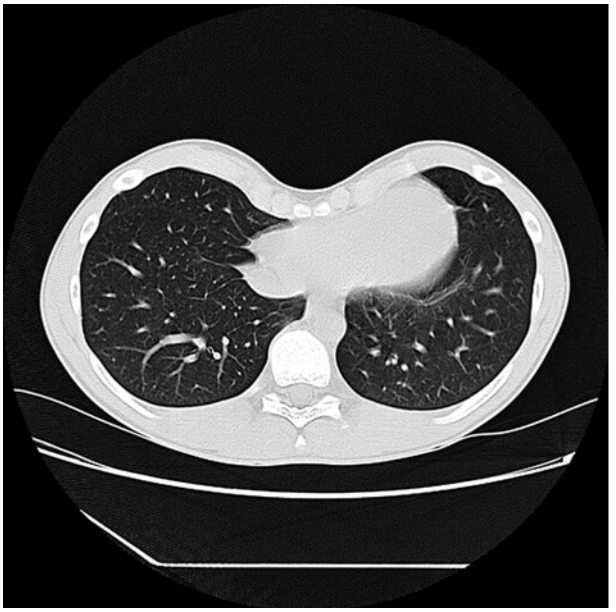

Fig. 4. CT scan of the same patient 2 min after evacuation of the vacuum bell

After this preliminary examination the further development and refinement of the vacuum bell treatment for improvement of funnel chest deformities started.

6.8.3 Vacuum bell procedure

All patients are informed that there exist different surgical options for funnel chest therapy, which could combine the requirements of aesthetic and functional remodeling of the thoracic wall. If patients decline an operation due to several or individual reasons, they are informed about the opportunity of a conservative therapy according to Eckart Klobe. They also are informed that this is a relatively new, however, not invasive kind of funnel chest therapy and that there are no long-term results available so far. If they still prefer this non-

Fig. 5. Application of the vacuum bell in the vertical position

surgical method, after standard clinical examination the optimal size of the vacuum bell is defined. The patients learn the proper application and utilization of the vacuum bell. First of all, it is important that the center of the polycarbonate window must be placed exactly above the bottom of the funnel. Usually the vacuum bell is applied in an upright position of the patient (Fig. 5). In patients with an asymmetric funnel chest deformity it could be useful also to apply the bell in a horizontal position. With the vacuum bell now being in position, the patients learn how to evacuate it. The patients are then equipped with the vacuum bell and start therapy at home for half an hour twice a day. After 1 month the patients present again as outpatients for consultation and discussion of potential problems with the current therapy. The position of the vacuum bell can be optimized and follow-up should take place now every 3 months.

It is suggested to start with a lower negative pressure during the first 3 months in order to allow the skin to adapt to the mechanical irritation caused by the foreign material that is applied with pressure and to relax the ligaments of the costal joints slowly. After that period the patients are requested to evacuate the bell with increasing power up to the maximum of vacuum, so that the skin under the polycarbonate window still shows sufficient blood circulation. After 3 months the patients should prolong the time of therapy up to 1 h, twice a day. The time of therapy should not exceed over 1 h twice a day because the skin due to the vacuum bell will suffer from this treatment irritation.

6.8.4 Patients

The vacuum bell treatment is feasible in patients of each group of age. The optimal age is between 9 and 18 years. In younger and smaller patients this treatment is applicable with the smallest bell designed for patients between the age of 5 and 8 years. The experience, however, shows that the treatment is not sufficiently effective in these small patients, because the skeletal structures of the anterior thoracic wall are still very pliable. As a consequence of this the funnel might sink back very fast into the initial position. It must be also discussed, whether such a therapy is useful in patients, who due to their youth do not yet care about the funnel chest deformity or as grown-ups do not mind about or suffer from their deformity. Especially these adolescent or adult patients might be stigmatized during the therapy and the formerly disregarded deformity becomes a psychological burden. Frequently parents wish this therapy for their children, because it represents an alternative to invasive surgery. It must be recognized, however, that also this vacuum bell therapy could be a painful and time-consuming treatment.

It is very important to ask the patients for their desires and expectations concerning this kind of therapy. If they doubt to have sufficient motivation to undergo such a therapy for up to or over 2 years, the vacuum bell treatment should not be suggested or performed. In some cases it could be useful, to test the vacuum bell treatment for 1 month, then interview the patient and reevaluate his motivation. For the case that ongoing compliance and motivation are absent or doubted for the vacuum bell treatment, it might be meaningful to discuss the variant of an operation.

The vacuum treatment is also feasible in older patients, but because of the increasing skeletal rigidity with age it requires more time to elevate the thoracic wall depression to acceptable almost normal condition and shape. During the last 6 years many patients of each age group underwent vacuum bell therapy for funnel chest deformity correction. All of these patients were very well motivated and compliant. The time of therapy averaged 30 months. After 1 month body posture improved markedly and the sternum depression could be elevated by 1 cm in all patients immediately after the vacuum bell application. After 3 months in 50% of the patients the elevation of the sunken sternum to a normal level

Fig. 6. Funnel chest patient before vacuum bell treatment and after 5 months of therapy

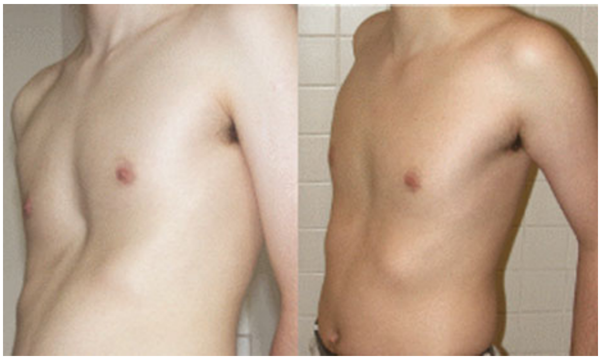

Fig. 7. Another funnel chest patient before vacuum bell treatment and after 5 months of therapy

lasted for 30 min up to 2 h, subsequent to daily therapy. During the following months an increasing consolidation of this status of elevation was obtained. Many of these patients complete therapy between 24 and 30 months because of achievement of a sufficiently aesthetic improvement and all of them were very satisfied with the results obtained (Figs. 6 and 7).

6.8.5 Complications

Complications in this therapy are very rare and harmless. There remains no permanent skin discoloration or discomfort. During initial treatment all patients experience moderate pain at the sternum and 53% of the patients report pain at the costovertebral joints. The pain, however, is very moderate, not requiring analgesic medication in any patient so far. One patient in our series suffered from unaccountable transient paresthesia in the right arm and leg during the first applications of the vacuum bell. Three patients experienced orthostatic disturbances during the first application of the vacuum bell, but this did not recur in subsequent procedures. In one patient we noticed dilatation of some skin vessels after 15 months of vacuum bell therapy. No other complications were observed.

6.8.6 Intraoperative use of the vacuum bell

The most difficult part of the minimally invasive repair of pectus excavatum (MIRPE after Donald Nuss) during insertion of the introducer and ensuing metal bar is to cross the space between the sternum and the pericardium to reach the other side of the thorax. This is because the space between sternum and pericardium is very narrowed in patients with a pectus excavatum deformity. Therefore it appears reasonable to reduce the risk of organ damage by enlargement of the retrosternal space using a tool to increase the safety of placing introducer and pectus-bar into the thoracic cavity. Some surgeons perform this intraoperative elevation using a bone hook through an extra skin incision at the sternum. To avoid such undesired incisions and resulting, occasionally hypertrophic or unsightly scars, it seems more useful to elevate the sternum or the anterior thoracic wall by non-invasive vacuum power.

During the Nuss procedure a medium-sized sterile vacuum bell is placed in such a way that the center of the window is placed right above the bottom of the funnel. The first steps of the Nuss procedure are performed in a standard way as described in Chapter 5.3.1.1. After preparation of the bilateral thoracic access for the ensuing intrathoracal part of the operation, the vacuum bell is placed exactly and evacuated under thoracoscopic view and the part of the anterior thoracic wall encompassing the sunken part is lifted. As long as the sternum is elevated, the placement of the introducer and the pectus-bar is much easier and with minimized danger of organ damage. It takes only 2 min to elevate the sternum, but it saves much more time with an easier and more secure placement of the hardware and with much better survey over the manipulation maneuvers within the thoracic cavity. Furthermore, based on this indirect rapid expansion of the thoracic cavity the thorax is preformed and the placement of the pectus-bar is much less traumatizing to thoracic wall structures in contrast to the conventional Nuss procedure (Figs. 8–10). This latter condition may be more the case for adult patients with a more rigid thoracic cage than for children.

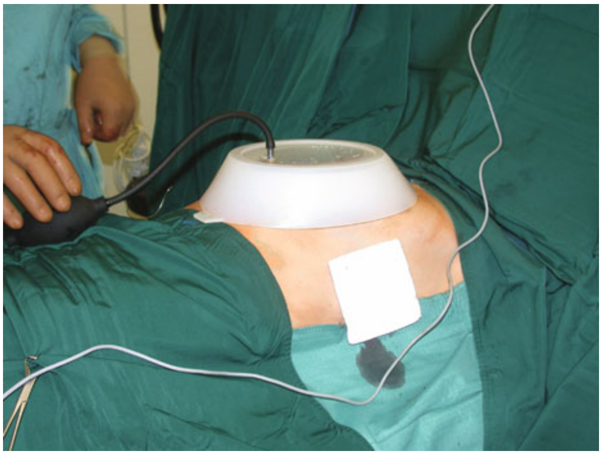

Fig. 8. Intraoperative application of the vacuum bell

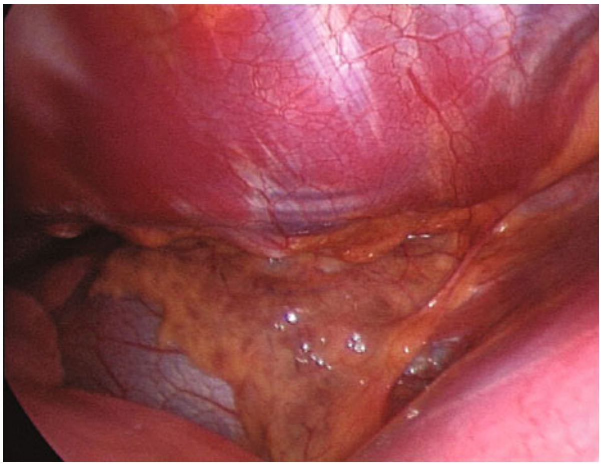

Fig. 9. Thoracoscopic view to the substernal space without the vacuum bell

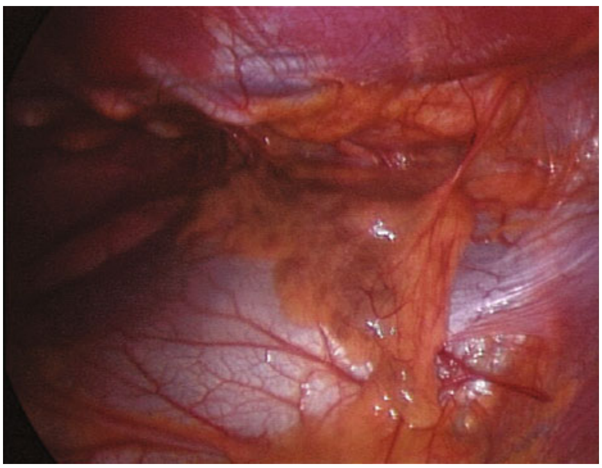

Fig. 10. Thoracoscopic view to the substernal space with evacuated vacuum bell

6.8.7 Preoperative use of the vacuum bell

There is an ongoing discussion whether to use the vacuum bell preoperatively prior to a Nuss procedure or not. It may become apparent that this kind of "pre-expansion" represents a helpful adjuvant pre-treatment for very deep funnel chest deformities. Especially in such patients, and because of the pre-lift of the sternum for several months preoperatively, the risks and pitfalls inherent within the Nuss procedure are reduced. The period of preoperative vacuum bell therapy depends on the deepness of the funnel bottom. Another discussion about preoperative vacuum bell therapy relates to the preoperative improvement of the body posture and the relaxation of the costal joint ligaments as well as intrathoracal muscles adherent to the sternum. This "pre-expansion" also seems to be advantageous to lower pain after the NUSS procedure, because this "pre-expansion" process loosens all anatomical structures withstanding the pressure of the pectus-bar.

6.8.8 Conclusion

The vacuum bell treatment according to Eckart Klobe seems to be a promising alternative for patients who decline surgical correction, for aesthetic indication as well as for functional improvement of impaired fitness. It also shows benefits in the pre-treatment of extensive deformities in order to facilitate the surgical access itself and thereby might also alleviate postoperative pain. However, there are still no long-term studies published and the reports on the application of the vacuum bell are small series with patient groups up to 100 patients so far, but the promising preliminary experiences raise expectations for further application and development.

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6.9 Pectus-bar removal technique

Anton H. Schwabegger

The technique of bar removal is shortly described already in Chapter 6.3 (Kuhn and Nuss) but herewith we intend to address some particular hints and technical requirements for at times intricate removal of the bar especially in the adolescent and adult patient, and furthermore if they have been left in place inadvertently for several years more than scheduled.

The type of anesthesia and the positioning of the patient do not deviate from the descriptions of Kuhn in Chapter 6.3 and ours in Chapter 5.2. The provision of current chest X-rays appears especially important for an appropriate localization of the metal parts. Such imaging is the more important, the more obese a patient presents, because herewith the pectus-bar wings and occasionally implanted stabilizers can hardly be found just by palpation underneath adipose and muscle tissue layers.

However, particular problems arise in removal of pectus-bars in adults, because the anatomy and shape of the thorax thus also the shape of the implanted pectus-bar present different and rigidity of the skeletal structures with different biological reactions, e.g., formation of callus, may complicate their removal. After duration of 3 years, the pectus-bar wings may also be dislocated to dorsal and thus the skin incisions along pre-existing scars from implantation must then be elongated further

Fig. 1. Incision at the left thoracal side, the end of the pectus-bar is hooked up with the flipper instrument, usually used to twist the pectus-bar during implantation. The pectus-bar wing is unbent with the flipper prior to extraction, in order to circumvent lesions of lungs and heart during extraction

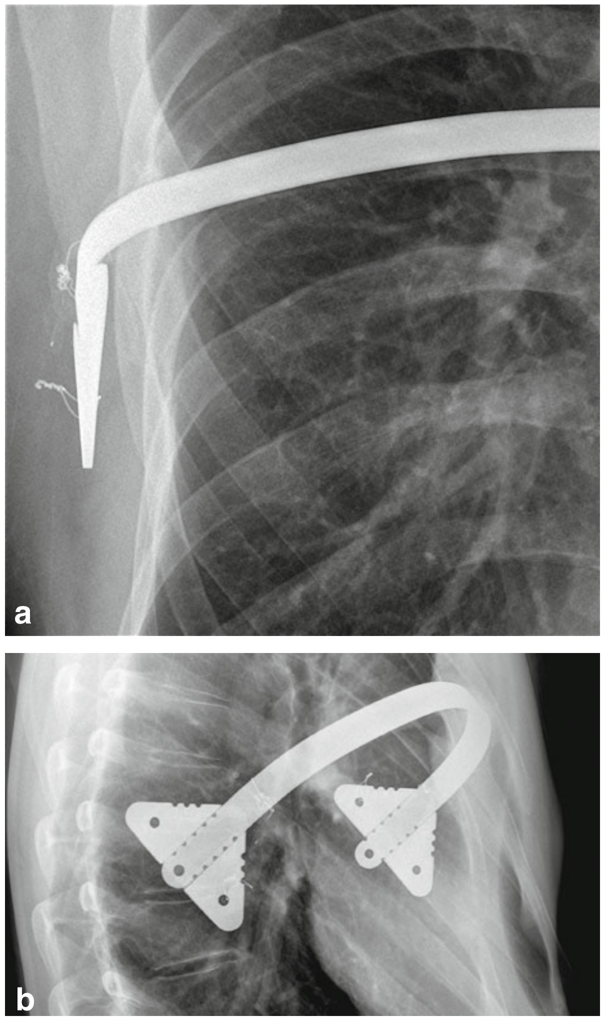

Fig. 2a. Right wing of a pectus-bar with attached triangular stabilisator, in a frontal X-ray aspect. Notice the broken fixation wires, potentially caused by dislocation of the pectus-bar wing and its stabilisator from the ribs. **b** Same situation in a lateral X-ray aspect. The twisting of the pectus-bar to cranially is clearly visible now, making the disruption of the wires understandable

to dorsal. This extension of the incisions may be necessary in order to mobilize the dislocated metal bar wings to that extent that it can be hooked to the pectus-bar flipper (Fig. 1).
In former cases however occasionally used but through dynamic respiration forces usually broken and therefore mostly dislocated fixation wires can be localized by means of the X-ray pictures prior to surgical incision (Fig. 2a, b).
In the case of major tipping of the pectus-bar it can at times become necessary to place the skin incisions, which serve for hardware explantation, remote from the pre-existing scars.
Occasionally used stabilizer plates must be shifted to dorsal in order to remove them from the bar wings prior to the pectus-bar explantation itself. This may be feasible only through extensive mobilization of the tissue embedding the bar wings and the adjoining part of bar strut, which wears the stabilizer. Herewith a sufficiently ample tissue pocket must be dissected and the adjacent, usually scarred musculature be mobilized to create sufficient place for the maneuver of mobilization of the stabilizer. A further problem arises in the use of triangular stabilizers, which for the positioning of the pectus-bar were promised to provide with more stability against the sequels of twisting. Then during scheduled explantation after years these triangular devices require much more ways to dorsal into the tissue for taking them off, which is up to two cm of space needed (Fig. 3a, b).
Removal of these stabilizers prior to pectus-bar removal is necessary to create sufficient space to place the flipper or similar tools to enable proper positioning for unbending the strut wings. Especially in the cases of adults, in which ongoing forced pressure, caused by the functional-anatomical factors (Chapter 2.1) weighing on the pectus-bar for several years, the bar wings are shifted further dorsally in many cases (Chapter 12.1). The originally set scars, however ideally placed for the access of hardware implantation, then no longer suffice for adequate mobilization of the stabilizers and bar wings, thus must be extended or placed further dorsally. Furthermore if formation of callus embedding the bar wings and stabilisators occurred after several years, positioning of the flipper instrument for bar unbending is cumbersome through a desired small still aesthetically pleasant incision (Fig. 4).

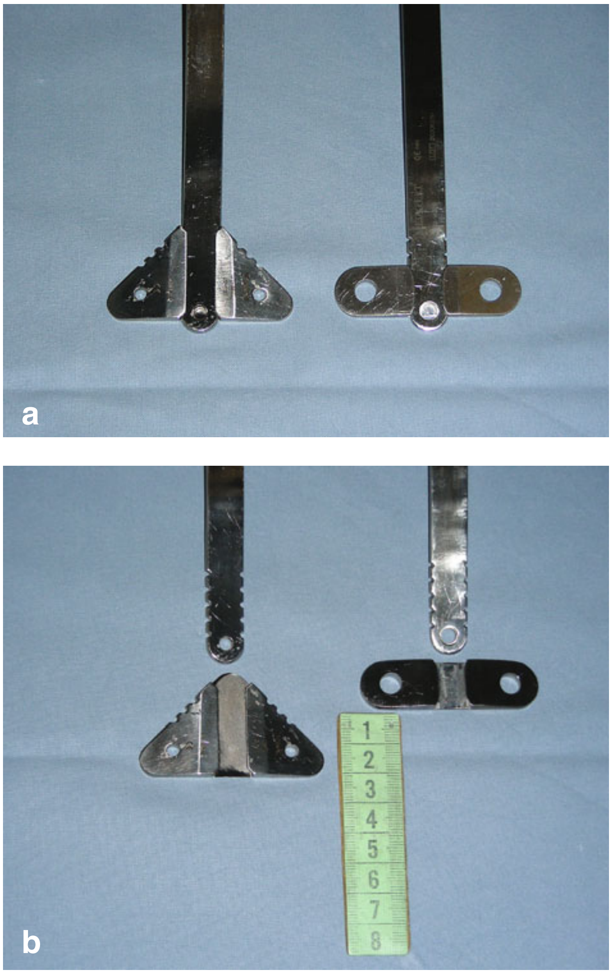

Fig. 3a. Triangular and strap-shaped stabilisators attached to pectus-bars to depict their size. **b** During explantation and detachment of the stabilisators this depiction gives evidence that the triangular stabilisator necessitates about 2 cm more range of posterior shifting along the strut to be shoven off

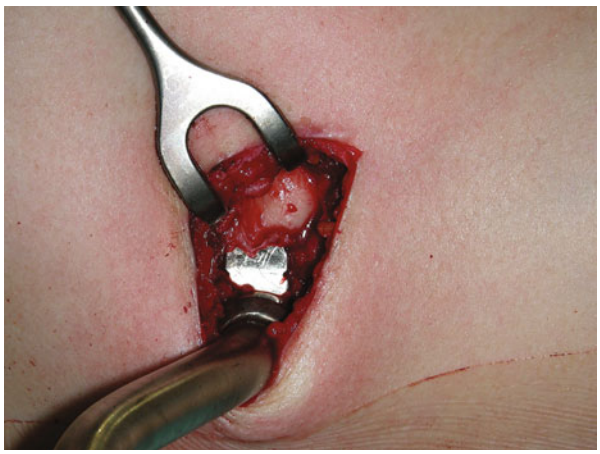

Fig. 4. A desired short skin incision may be of disadvantage, if bony callus formation cuffs the pectus-bar, which renders positioning of the flipper instrument for the purpose of unbending the wing of the strut difficult

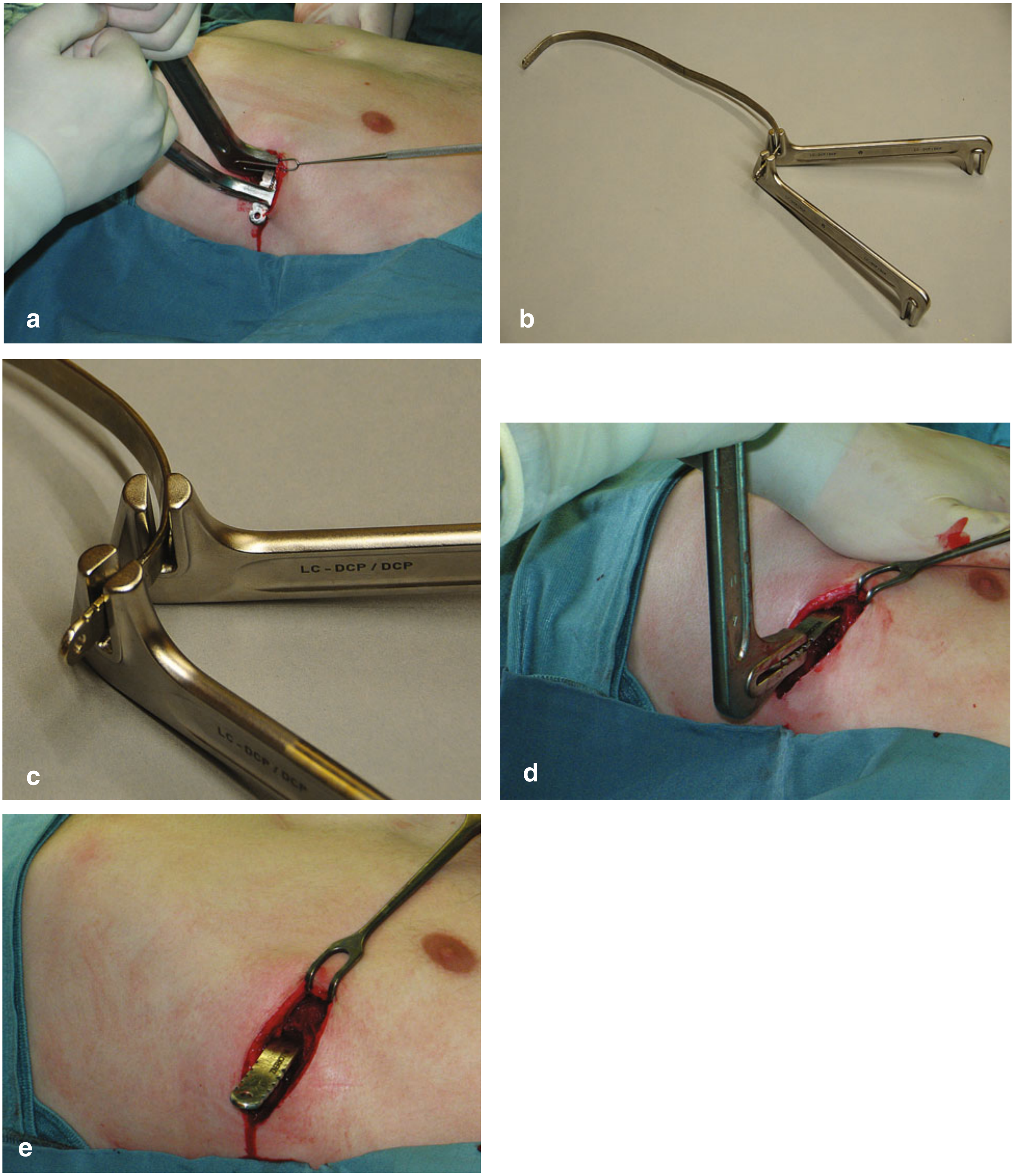

Fig. 5a. Wrench irons set in place to easily unbend the left wing of a pectus-bar into a horizontal plane, in order to enable extraction from the right side. **b** Wrench irons placed at a pectus-bar demonstrate the ideal site of application for proper unbending in order to circumvent rib fractures. **c** Close-up view to show the ample jaws of the wrenches that may be advantageous in cases, where the wings of a pectus-bar are already curved or damaged to such an extent, that the slot of the flipper instrument usually well fitting to the bar wing may not be attached any more. **d** The wrench at its second end is constructed with jaws wider than the other one, which are able to embody even a distorted wing of a pectus-bar for adequate unbending. **e** Pectus-bar wing bent into a horizontal plane, ready for securing extraction from the other (right) side. Sterile gel applied to this end may alleviate the transthoracal extraction

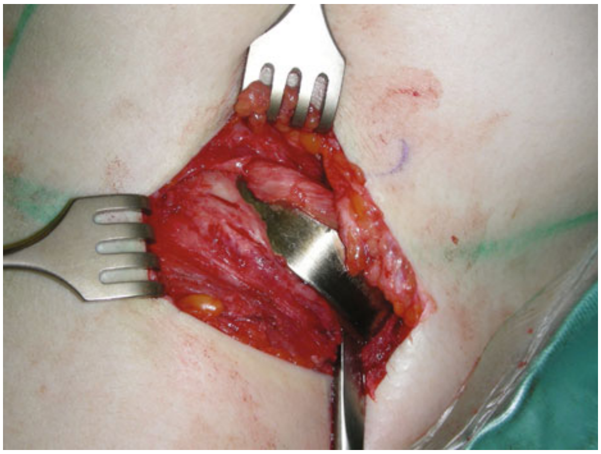

Fig. 6. Chisel needed to liberate pectus-bar embedded in bony callus formation

In such cases, when the flipper instrument cannot hook up the bar wings, wrench irons, usually used in osteosynthetic trauma surgery, may be helpful to lift up and simultaneously bend the bar wing to a desired position (Fig. 5a–e).

Prior to bending up the bar wings, which in some cases are embedded by bony callus tissue, such excess of bone must be removed using sharp chisels (Fig. 6), in order to avoid rib fractures resulting from undue bending forces. Anyhow, if such callus is present, it should be removed entirely, because after pectus-bar removal such callus remnants may generate new callus due to the irritation of surgical manipulation and may cause complaints such as tenderness on pressure, visibility and even aesthetically eye-catching humps in slim patients. Occasionally during growth spurts and through respiration movements friction between the pectus-bar and the bony part of the rib develop because of increasing tight contact. Thus bony erosions (Fig. 7) and weak zones at the rib exist, which might result into a painful fracture, if inappropriate forces are applied during pectus-bar explantation. In order to avoid such complications it is a prerequisite to have actual X-rays of the chest present prior to perform explantation surgery.

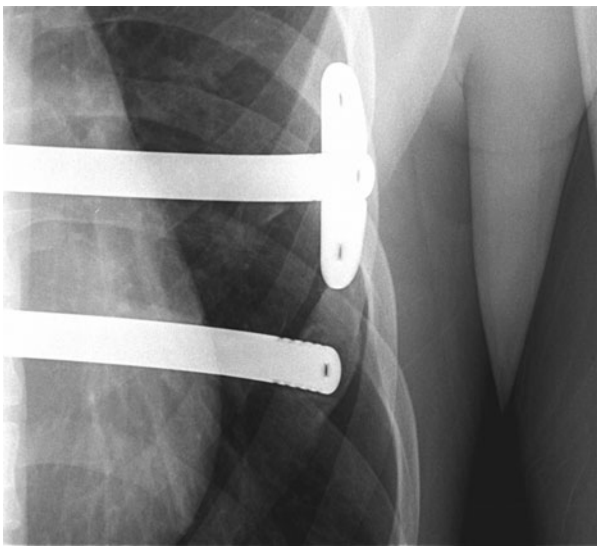

Fig. 7. Erosion of bone at the wing's end of the lower pectus-bar caused by ongoing friction and pressure due to growth spurt and permanent respiration movements

It appears very meaningful, that already during the pectus-bar implantation on the one hand the size of the pectus-bar is selected not too long, and that the skin incision is not set too far ventrally on the other hand. However, nowadays we do not find the utilization of laterally placed stabilizers very useful, as the wings of the pectus-bar can be sufficiently fixed to the ribs using absorbable strong sutures and because of other distinct reasons [1], which are described in Chapter 6.4.

At our institution, because of the prevalence of adolescents and adults to be operated, the pectus-bar explantation is carried out exclusively in supine position. Some authors sometimes describe it to be performed in lateral decubitus position. This maneuver only then succeeds without danger, as long as the tips of the metal strut are only slightly bent up and removal along the radius of the arch of the pectus-bar as well as of the chest is feasible. That is only the case, if the pectus-bar lies in situ almost perfectly round-arched. However, such an arched form of the pectus-bar almost exclusively only in children and therein very elastic thoracic cages is

Fig. 8. Comparison of two shapes of the pectus-bar used at different ages. The right (upper) one is shaped like a wave or "M" and is used frequently in adolescents and adults with a matured shape of the chest. The left (lower) and smaller one is almost curvilinear round and formed for implantation in children with yet pliable skeletal structures

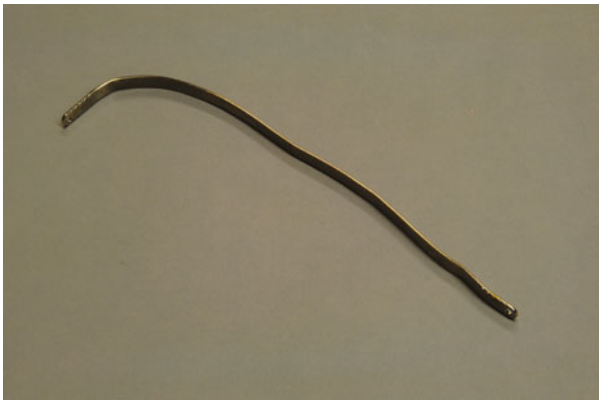

Fig. 9. Pectus-bar extracted from an adult patient, showing one wing untreated, while the other wing that passed through the chest cavity is bent up to a horizontal plane to avoid any intrathoracal injuries

used (Fig. 8). Upon hardware extraction then the thoracic wall may follow the shape of the pectus-bar due to its elasticity without any problems, but it will not behave like that in elder patients with a rigid chest. In adolescent or adult patients, this round-arched shape of a pectus-bar never comes into consideration, because at that age the transversal diameters and the anterior thoracic wall are almost always shaped like a horizontal "8" thus the implantation of a round-arched strut would be impossible or in danger to twist or dislocate because of undue pressure. On the basis of this substantially different overall shape of the thorax, a differentially formed pectus-bar is required to lift up a sunken sternum and the adjacent ribs. The metal strut in these cases is formed like an undulatory "M" (Fig. 8), thus in contrast to children may not be extracted along a simple curvilinear round arch in lateral decubitus position. Intentional round extraction without bending the distal end will definitely hook up inner organs with resulting fatal consequences. Prior to extraction, one end must be unbent almost flatly into a horizontal plane (Figs. 5e and 9) in order to avoid such injury to the intrathoracal organs (Chapter 6.3).

On the basis of capsule development around every implanted foreign material, a rigid scar will also cover the pectus-bar even intrathoracally after months and years. That is why especially in adult patients the pectus-bar seems to be trapped by that scar tissue, which renders extraction of long struts rather difficult. It may be helpful in these cases, to hook up two bar flippers at the strut, each at one side of the unbent wing. Then along the axis of the straightened bar rotation is applied with force to loosen the intrathoracal capsule.

In children with a short length of the pectus-bar this is of fewer importance, but is necessary in adults with very long struts, especially if the bar stayed in situ for several years and is shaped with multiple arches. The distal and bent wing of the pectus-bar is moistened with sterile gel to alleviate its extraction and to circumvent any traction forces of the interdigitations at the bar's wing within the intrathoracal scar-capsule.

After explantation the patients are followed-up for another year to observe the further course, taking photos and measurements with thorax calipers.

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Special techniques in the keel chest deformity

7

7.1 The Ravitch procedure

Anton H. Schwabegger

The etiology of the keel chest deformity, still being a hypothetical explanation and a matter of investigation, is probably caused by secondary elongation of rib cartilages during growth spurts, which result in prominences of the rib themselves or are conjoined with a protrusion of the sternum (Figs. 1 and 2a–c) to a variable extent and shape. Therefore, the surgical correction is aimed at shortening the affected cartilages with or without sternum osteotomies.
More rarely such cartilaginous prominences remain isolated from a sternal protrusion, which means that the sternum itself is located in an anatomically orthotopic position, while the adjacent rib cartilages bulge forward symmetrically and appear as parasternal crests extending along a longitudinal axis (Fig. 10 in Chapter 2.3.2). If such cartilaginous crests are present solely unilaterally (Fig. 9 in Chapter 2.3.2), the sternum is usually rotated to the concomitant side, challenging the surgical repair with respect to gain sufficient symmetry.

Fig. 1. Lateral view of X-ray in a male with chondrogladiolar pectus carinatum deformity

7.1.1 Surgical technique

Since the development of the fundamental basics of the surgical technique of pectus carinatum deformity, only minor variants have been described. The basic surgical steps were developed by Robicsek 1963, Welch 1973, Fonkalsrud 2001, and several others [2, 8, 14] and finally resulted in the integrative concept summarized by Ravitch [7], which due to its convincing outcome lasts for decades now [1, 3, 4, 9–12, 14]. Only minor modifications since then are developed, exemplarily noticed the redressing with sternal struts [3] or the muscle split technique [13] to avoid invasive muscle flap elevation (Chapter 7.2).
Usually a midline incision at the sternum enables sufficient survey at the site of remodeling (Fig. 3). Occasionally in unilateral deformities a horizontal incision or in females incisions along the inframammary creases may be opportune (Chapter 5.1).
Prior to the access to the cartilages and sternum in the Ravitch technique however, the Pectoralis major muscles are dissected free and elevated from their sternal and inferior rib insertions to expose the skeletal structures that have to be shaped. The Rectus abdominis muscles may be detached from their origin at the lower ribs, interdigitating with the Pectoralis major muscles. This detachment is performed with inclusion of the xiphoid bone, which means that the xiphoid remains attached thus vascularized via the muscles. The main purpose of such dissection of the xiphoid consists therein that the xiphoid and the muscle origins may more easily be reaffixed to the sternum with strong suture or wires *than* a solely cut muscle without sufficiently strong tissue parts then at the final step of remodeling.

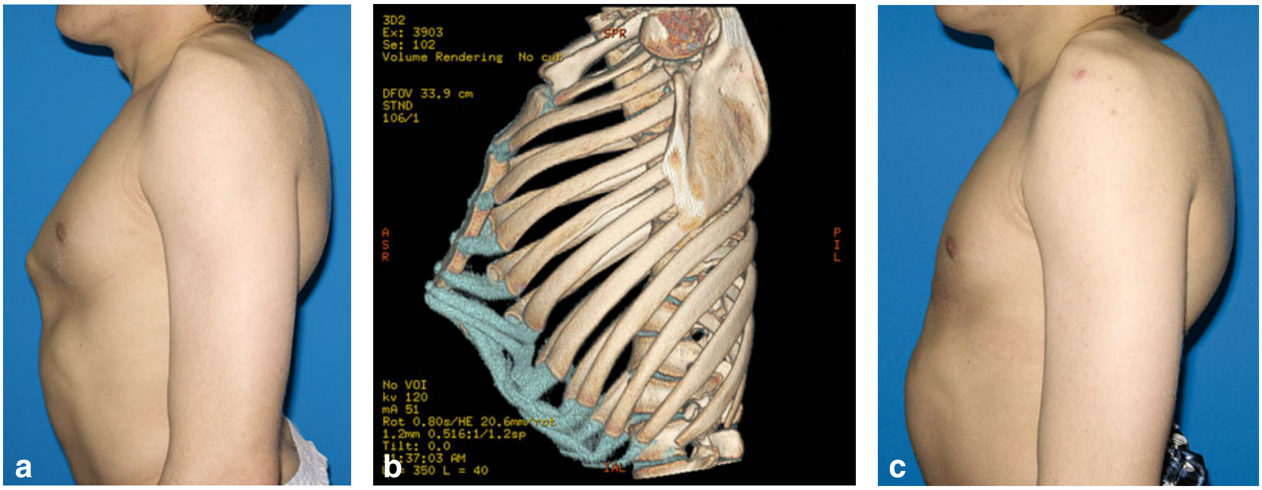

Fig. 2a–c. 20-Year-old male with osteodystrophic microsomia and an extensive chondrogladiolar keel chest preopreatively (**a**) and the corresponding CT-scan depicting the extent of deformity (**b**). Postoperative view with very pleasing aesthetic result

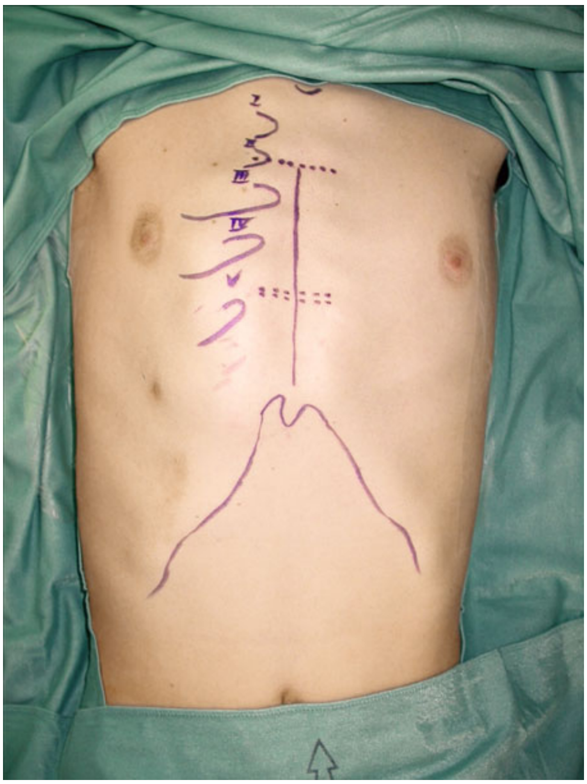

Fig. 3. Intraoperative planning of incision (vertical midline) in 17-year-old male with unilateral keel chest deformity predominantly at the right side. The sites of osteotomies are outlined with short horizontal markings

Fig. 4. Cartilages resected subperichondrally from the 3rd to 6th rib in a case with a bilateral deformity

The first basic step however consists of multiple parasternal rib cartilage resections for the purpose of shortening overabundant length whether they are deformed in a convex or concave manner. Some of these cartilage incisions (chondrectomies) may solely be performed as chondrotomies, when appropriate length of single ribs at individual levels is present. At each individual level with cartilage excess, chondrectomies are done

(Fig. 4), whereas at other levels only a weakening of a distorted cartilage may be advantageous. The extent of cartilage resection in a still growing child should be limited to what is absolutely necessary [3] in order to minimize postoperative development of heavy scarring, which might result in a cumbersome adverse effect, an acquired Jeune syndrome with severe pulmonal restriction based on an anterior thoracic wall growth restriction [5, 6].

The particular characteristic in doing such chondrectomies consists in subperichondrial resections, which means that the cartilages are "peeled" out from the perichondrium tubes, the access managed by door-like incisions at the anterior surface of the ribs. By this, the perichondrium tubes remain in situ for further availability for restabilization. Furthermore, by the subperichondrial access any lesion to the pleura and the intercostal nerves is kept to a minimum.

The second basic component of this concept is single or multiple horizontal sternotomies (Fig. 5) at variable levels, depending on the kind of deformity, whether a chondromanubrial or a chondrogladiolar deformity. Furthermore, even asymmetric cases follow these prin-

Fig. 5. Lateral chest X-ray depicts the situation postoperatively with three horizontal osteotomies performed for remodeling. The lowermost gap represents no osteotomy but a not yet ossificated growth zone, already resulting in a tissue protrusion several months postoperatively

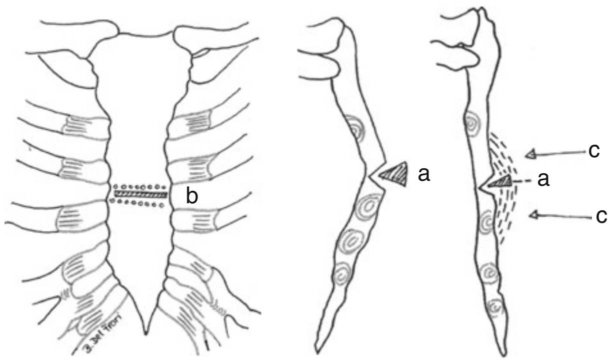

Fig. 6. Schematic depiction of wedge resections (*a*) or horizontal osteotomies (*b*) at the sternum, the level of singular or multiple incisions may differ depending on the shape of deformity and aimed remodeling. A bony wedge may even be used as a transposed autograft (*a*) in order to fill up the defect, which is produced by unbending (*c*) an incised part of the sternum in order to support its final shape

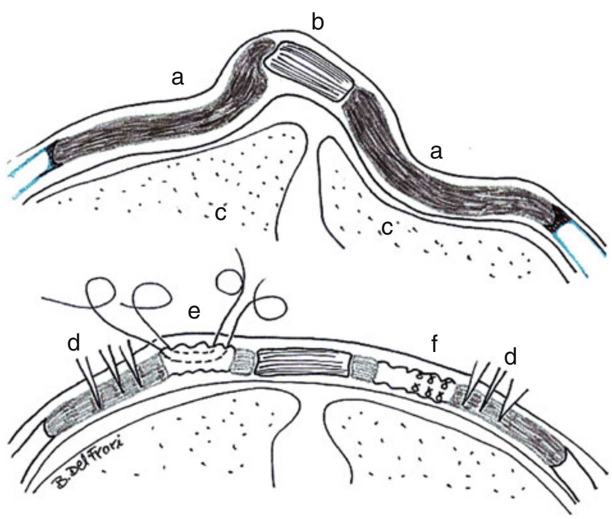

Fig. 7. Deformed cartilages with abundant length (*a*) will be partially resected and for the case of sternum malrotation (*b*) a horizontal osteotomy must be performed. The lungs (*c*) will not suffer from such a remodeling procedure with neglectable alteration onto the intrathoracic volume. The remaining rib cartilages, if still showing protrusion, may be incised at single or multiple sites (*d*) to allow unbending and giving way to the shortening effects on the perichondrium tubes by reefing sutures (*e*, *f*)

ciples. Sternotomy ideally is performed near or at a major protrusion as a wedge resection osteotomy on the one hand, or an incision creating a gap along an ascending slope of the sternal bone in order to enable redressing into a frontal plane (Fig. 6). Usually only the outer table of bone is incised, whereas the inner table will be gently green-stick fractured by forced manual manipulation. If an asymmetric deformity with malrotation of the sternum is apparent, the osteotomy must also include the entire thickness of the sternal bone, in order to alleviate its derotation into a physiologic plane. Incomplete transsection in the symmetrically protruding sternum avoids further dislocation or disruption with the potential sequel of painful sternum pseudarthrosis. If a complete osteotomy for sternum derotation is mandatory, the site of osteotomy therefore must be refixed with appropriate materials.

The third step then is the remodeling process specially featured by perichondrium tube reefing sutures in order to shorten the exceeding surface of the anterior thoracic wall and to restabilize it (Fig. 7).

Thereafter the detached xiphoid with adjacent rectus abdominis muscles will be refixed to the down mobilized and at occasion slightly shortened sternum to put it under tension, thus supporting its flat fixation within a frontal plane [8]. In the case of a present gap produced by osteotomy of the sternum and its redressing to posterior, such a gap may be filled with a bony wedge to allow proper healing there. Every resected part of bone may be used to splint another gap created by the

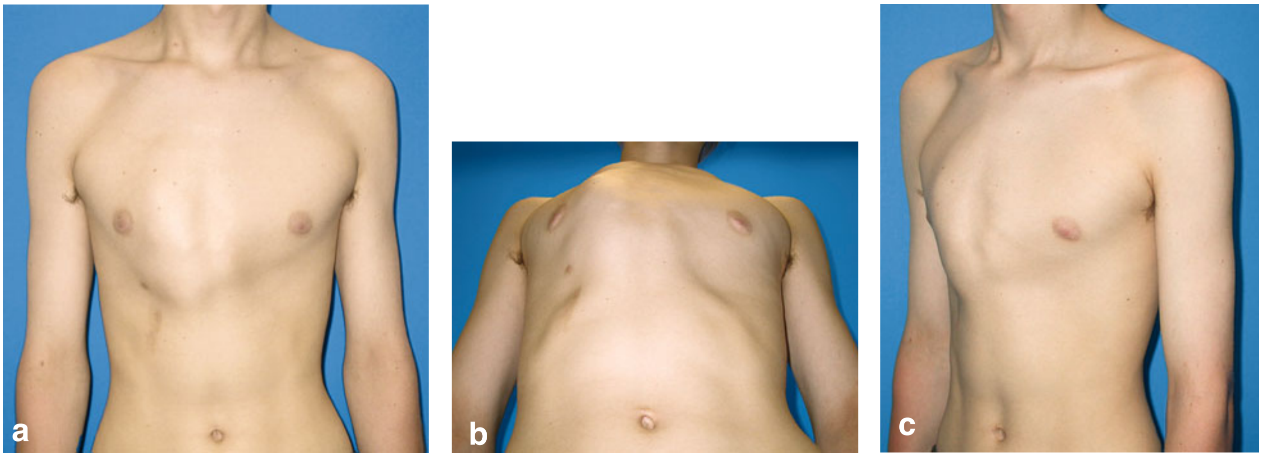

Fig. 8a–c. Same patient as in Fig. 3, preoperatively

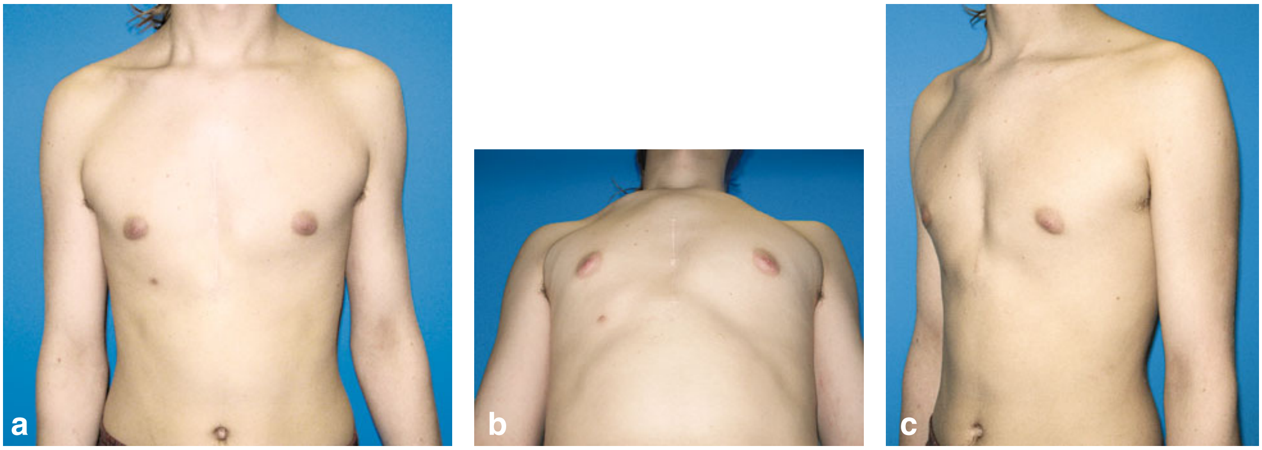

Fig. 9a–c. Same patient 1 year after pectus carinatum repair following the principles of Ravitch, but modified with a 2 months period of postoperative bracing

bone remodeling procedure (Fig. 7). The pectoralis muscle flaps are anatomically repositioned to the sternum and the ribs to regain and maintain their original function.

In general, these steps of the basic concept after Ravitch may be applied to the majority of all variants of keel chest deformities (Figs. 8 and 9), since the key points of shaping all kinds of protrusions consist of redressement of mobilized bone and rigid shortening of lengthy ribs. Usually the aesthetic outcome of this kind of surgery even applying several modifications results in good to excellent appearance in up to 97% of all patients treated for pectus carinatum deformities [3].

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7.2 Modifications of the Ravitch technique for correction of pectus carinatum with split muscle, bioabsorbable osteosynthetic material, and brace: the Innsbruck concept

Anton H. Schwabegger, Barbara Del Frari

7.2.1 Introduction

In this chapter, the experience with a modified access for the correction of the pectus carinatum deformity is described. The modifications versus the classic Ravitch [19] procedure exist in multiple minor splitting of the pectoralis major and rectus abdominis muscles [25], the use of bioabsorbable osteosynthetic material and the application of a compression brace postoperatively [24]. Especially this combined treatment access has proved itself very advantageous, on the one hand to reduce pains and on the other hand to improve the late result in regard to shaping and minimizing scar markedness.

7.2.2 Methods

First, especially the use of the muscle split technique permits mobilization of the shoulder belt relatively early after surgery, because in this access the pectoralis major and the rectus abdominis muscles are solely split along their fiber direction and in contrast to the conventional Ravitch technique need not to be elevated as partial muscle flaps from the ribs and the sternum.
Second, as an additional modification, and opposite to earlier described methods we nowadays use bioabsorbable plates and screws (Bionx, Tampere, Finland) [2, 5, 12] for refixation of the osteotomized sternum, which disintegrate after one to 2 years completely.
Third, to improve the long-term result of remodeling and stable healing of the sternum osteotomy and close-by chondrotomies as well as to reduce the risk of hypertrophic scar formation, a special custom-made keel chest brace (Chapter 7.4) is applied for 2 months [24, 25].

7.2.2.1 The muscle split technique

The conventional access to the correction of the pectus carinatum was described in many series and sufficiently long-term observations over the past decades. Above all the authors Ravitch, Robicsek, Fonkalsrud, and several others have contributed very much to the previous development and refinement of surgical development [4, 6–8, 19–22, 26]. The refinements of these authors predominantly range on modifications of the osteochondrotomies and fewer on the treatment of the soft tissue. However, precisely because rehabilitation time [23] is substantially linked with function of the pectoralis major and rectus abdominis muscles, we considered how the postoperative mobilization and healing phase could be reduced with regard to impairment of the surgical damage to the muscles. In the conventional technique, the sternal and partially the costal part of the pectoralis major as well as the cranial part of the rectus abdominis muscles are detached from their periostal insertions to the sternum, xiphoid and ribs and then elevated to expose the relevant rib cartilages (Fig. 1). This procedure up to date appeared to be necessary to alleviate a clear access to the site of osteochondro-

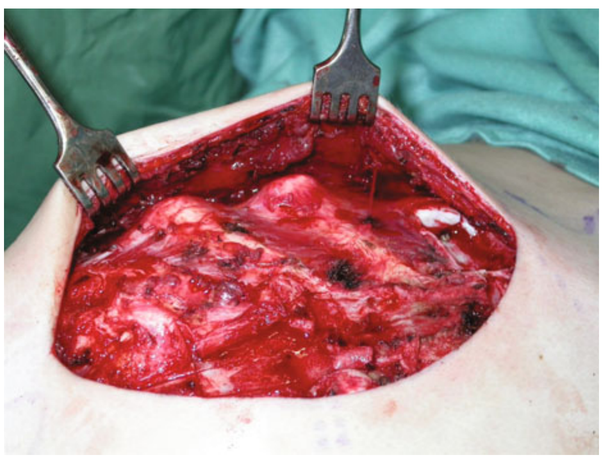

Fig. 1. Skin and muscle flaps elevated to provide with access to the deformed rib cartilages, the intercostal parasternal perforator vessels are transected

tomies. This rather invasive procedure with raising of muscle flaps on the one hand is of major advantage because of the outcropping survey to the anatomical structures to be dissected and on the other hand it also permits a very exact judgment of the surgical steps of remodeling the anterior thoracic wall.

Apart from that issue these muscles are developed very vigorously particularly in young athletic male patients and must be refixed anatomically with outstanding accuracy, what is to be managed at the sternum and periosteum sometimes very badly. The edges of the sternal part of the pectoralis major muscle may be easily sutured together in the median line, but this action will create an unnaturally looking aesthetic appearance of the décolleté. Furthermore, it is of disadvantage that this anatomically reattached muscles, based on the powerful lever effect of the shoulder belt, must be put to rest and be partially immobilized for up to 6 weeks. With regard to postoperative rehabilitation such a period of physical exercise restriction clearly affects the quality of life temporarily and should be subject of avoidance [25].

It therefore appeared meaningful to elaborate a method, with which the muscles did not need to be elevated and thus temporarily suspended from function. A transmuscular access then seemed to be the natural alternative, for each rib individually and with only minor damage to the muscles coevally. In 1970 Jensen [11] described such an access with several transmuscular incisions to reach the individual ribs for transection, however in the pectus excavatum deformity. Likewise these transmuscular accesses in the correction of pectus carinatum are aligned (Figs. 2–6) along the muscle fiber direction and therefore do not substantially damage the muscle and further on only temporarily and slightly impair the entire function. The sternum osteotomy in most

Fig. 2. Marked blue line at the Pectoralis major muscle showing the incision line along the muscle fibers for the transmuscular access to the ribs

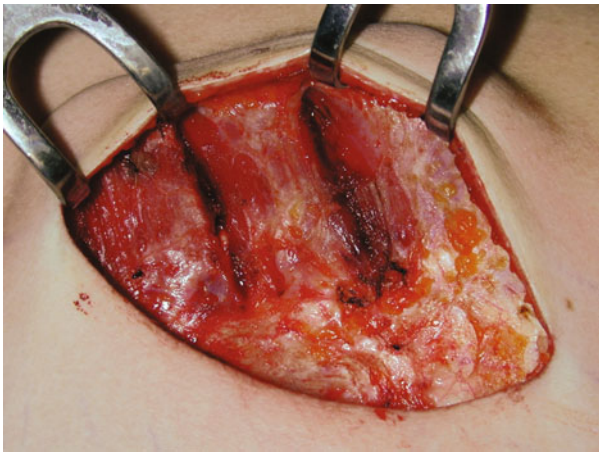

Fig. 3. Individual incisions through the muscle fascia and ensuing blunt separation of muscle fibers depict the ribs underneath with only minor damage to the muscle itself

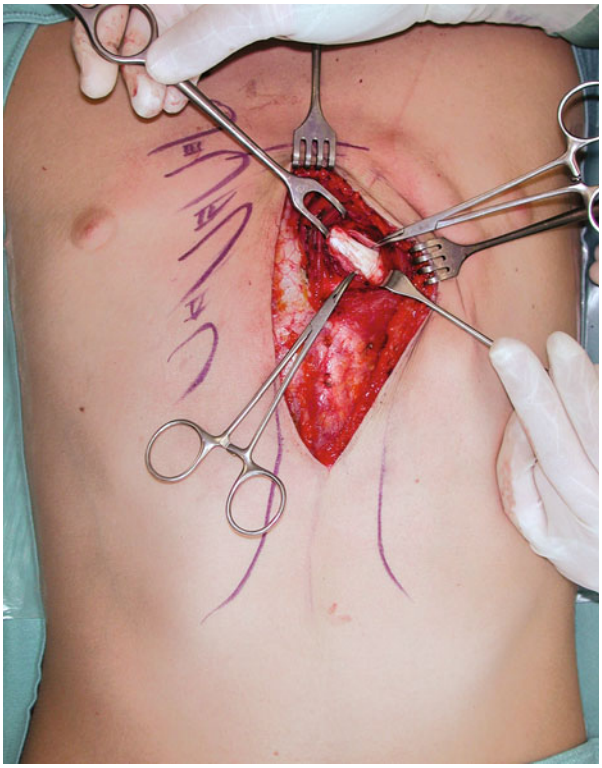

Fig. 4. The muscle margins are retracted and the individual ribs exposed for sub-perichondral resection

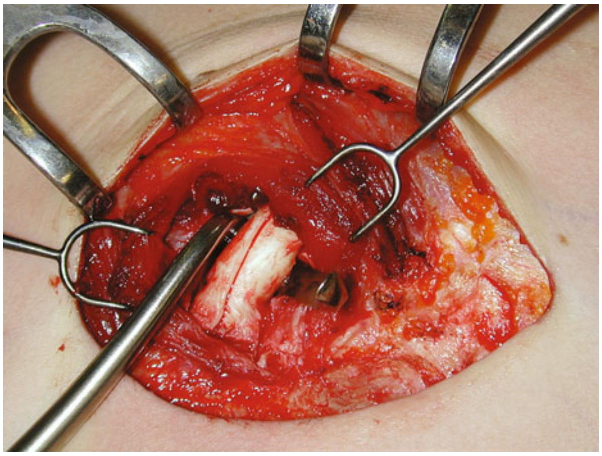

Fig. 5. A specially shaped rib dissector is used to elevate the rib cartilage and enables its careful transmuscular resection

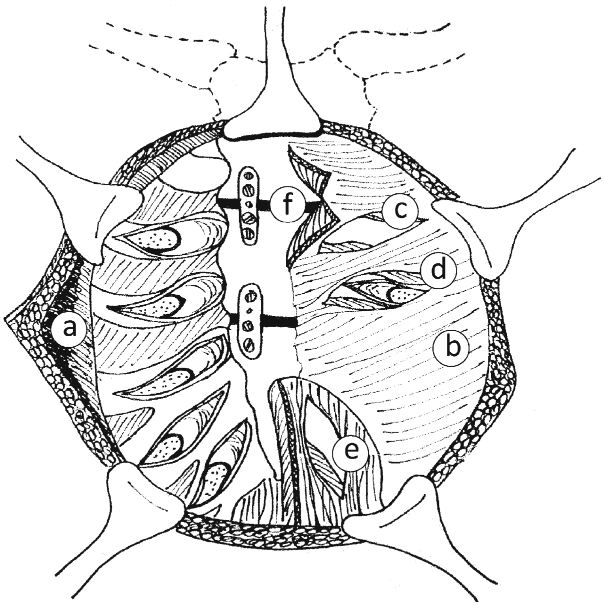

Fig. 6. Schematic overview of conventional technique at the patient's right side and muscle split technique at the left side. *a* Retracted pectoralis major muscle and skin flap with rib cartilages resected. *b* Pectoralis major muscle left in place with several transmuscular splittings (*c* and *d*), *e* muscle split along the rectus abdominis muscle to expose the seventh and eighth rib cartilages, *f* slightly elevated periosteum and horizontal sternum osteotomy with absorbable osteosynthetic material

cases necessarily along a horizontal line usually can be carried out (Fig. 7) easily by a separate access with a door-like incision (Fig. 8) of the periosteum without essential relief of the adjacent pectoralis major muscles. After osteosynthesis with bioabsorbable plates and screws (Figs. 9 and 10) the elevated periosteum

Fig. 7. Planning of surgery with skin markings showing the site of vertical median skin incision and horizontal sternum ostetomy

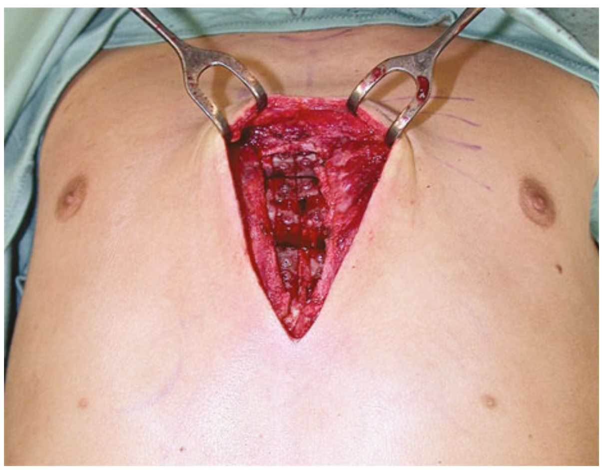

Fig. 8. Wing-door shaped incision of the sternum periosteum, the absorbable plates and screws bridging and fixing two horizontal osteotomies. Note the muscle margins are still inserting to the lateral sternum borders

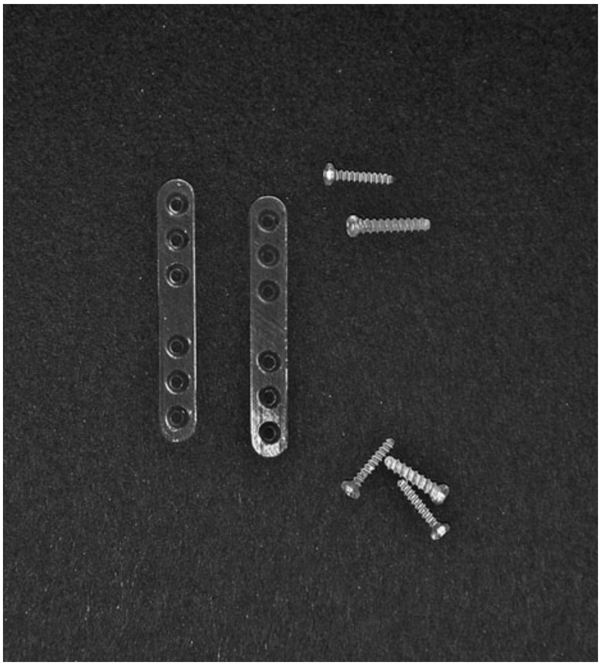

Fig. 9. Absorbable osteosynthetic material used for sternum refixation

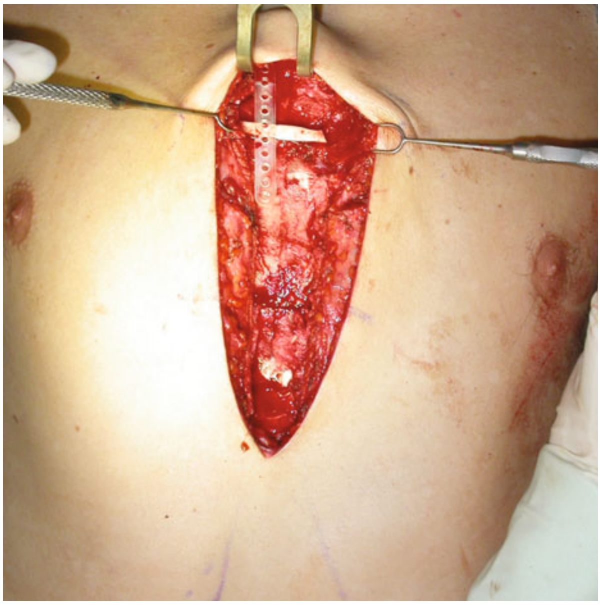

Fig. 10. The wedge-shaped horizontal bone incision is filled with a cortical bone part excised from the lower anterior compacta of the sternum, fixed, and kept in place by the absorbable plate

will be reattached above the embedded osteosynthetic parts with strong sutures. In minor, unilateral or also in more extensive keel chest deformities this

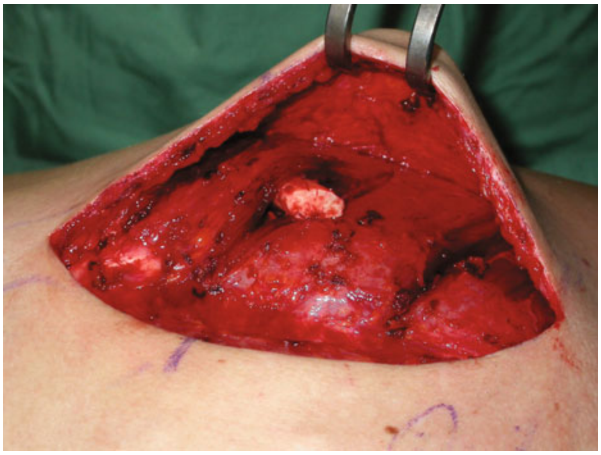

Fig. 11. Transmuscular dissection of rib cartilage in an athletic male with considerable pectoralis major muscle bulk

muscle split technique has proved itself exceptionally helpful [25]. The operation times do not substantially exceed those of the conventional technique, however, due to the retromuscular depth of the location of ribs (Fig. 11), the overview for the sub-perichondral resection of rib cartilages is somewhat restricted. In doing so, the risk of pleura lesion is probable. Based on that narrowed survey, above all in very athletic patients with thick muscle bulk, this access appears somewhat complicated. On the other hand, right in athletic persons with the urge to the muscle exercises and sports activities, a long period of movement and strain restriction will result in inadvertent atrophy of well-defined muscles.

7.2.2.2 Surgical technique

The skin incision design does not differ from the classic Ravitch procedure, usually a cranial to caudal vertical cut along the median line is performed (Fig. 7). In female patients, the incisions are set preferably along the submammary crease (Fig. 12) not to compromise the décolleté in its appearance (Chapter 9). Also in solely unilateral deformities, in which under circumstances no sternum osteotomy is necessary, the skin incision may be orientated horizontally (Fig. 13, Chapter 5.1). Subsequently the skin and subkutis flaps are elevated along the pectoralis major and rectus abdominis muscles (Fig. 2). The ribs of interest are palpated through the muscles and the muscle fibers are bluntly separated to reach the perichondrium of the ribs (Fig. 3). These transmuscular accesses measure 3–6 cm each depending upon the length required for rib cartilage transection or resection. Herewith an individual transmuscular access is created

Fig. 12. Surgical access is a female with asymmetric and predominantly unilateral pectus carinatum deformity, using a submammary incision only

Fig. 13. Horizontal skin incision in a male with unilateral deformity. All deformed ribs cranial and caudal from such a skin incision may be reached for excision or transection

for every rib cartilage (Fig. 5). In areas where the rib cartilages course more oblique to caudal, one muscle separation slit allows access to even two adjoining rib cartilages. This is rather probable at the level of the fifth, sixth, and seventh rib, where the rib cartilages are joined and a correspondingly widened access must be selected. Above all, it is of importance to split muscles along the direction of fibers as well as the insertion fascias preferably in a rectangular direction to the ribs in such a way that almost all of the muscle insertions remain in situ. Because at the lower ribs, where cartilage parts are transected and/or resected, several muscle slips from the oblique abdominal muscles insert. Therefore it is of paramount importance to put accurate reefing sutures (Chapter 7.1) to the perichondrium sleeves to guarantee stable muscle function. Furthermore all of the anterior fascia sheet of the rectus abdominis muscle must be repositioned and secured tightly with sturdy sutures in order to avoid abdominal wall bulging, which however is more common in the conventional procedure with total muscle release from the lower ribs and xiphoid [14, 23].

The length of the muscle separation should not exceed 6 cm in order not to impair the muscle innervation, as the concerning nerves course from lateral to medial. Emerging from the brachial plexus as medial and lateral pectoral branches they initially, at the lateral third of the muscle portion and underneath of it, run in a vertical fashion. Subsequently they divide further up in tiny branches, align to the muscle fibers, and enter them [3, 15, 18, 28]. The more medial blunt muscle separation for access to the cartilages is performed, the lesser is the probability of innervation damage (Fig. 14).

A special additional advantage using this muscle split technique without elevation of muscle flaps consists therein that the parasternal perforator vessels emerging from the Internal thoracic vessels remain pre-

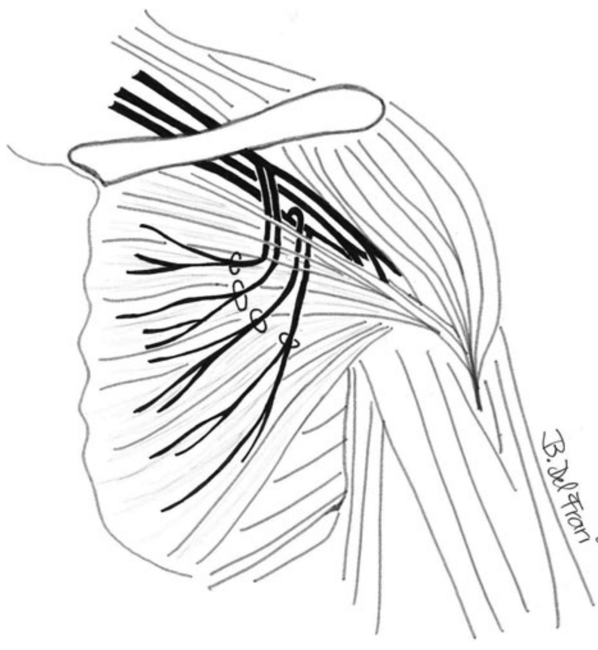

Fig. 14. Schematic depiction of the medial and lateral pectoral motor nerve branches emerging from the brachial plexus. Note the course, first descending vertically and then more medially running parallel to the muscle fibers, where blunt muscle splitting is performed to expose the rib cartilages

served totally. This fact of entirely untouched and preserved muscle vascularization at the parasternal region enables an optimal wound healing at the area of thoracic wall remodeling. It will obviously and substantially allow a quicker healing in contrast to the procedures, in which the muscles are elevated as flaps and many perforator vessels have to be transected inevitably.
For the case that in females a horizontal sternum osteotomy has to be carried out, this usually succeeds also from the incision along the submammary crease. Herein the utilization of an angled retractor with attached light source (Fig. 15) enables sufficient overview underneath the elevated skin flaps. Furthermore, an oscillating saw with an angulated blade has proved itself as very advantageous to reach the distantly and subcutaneously localized bone transection line (Chapter 5.1, Fig. 16).

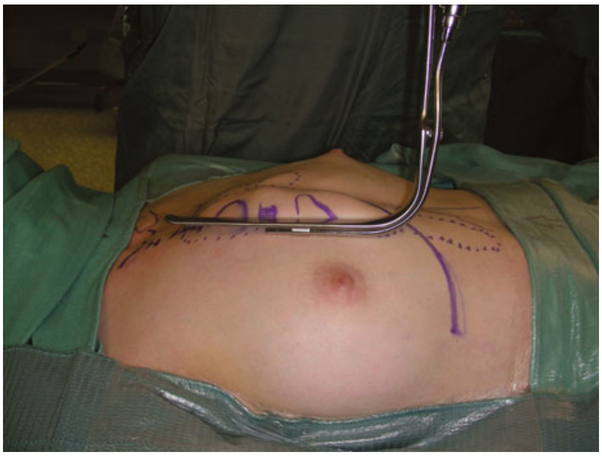

Fig. 15. An angled retractor equipped with a light source to allow appropriate overview for surgical maneuvers beneath the elevated skin flap

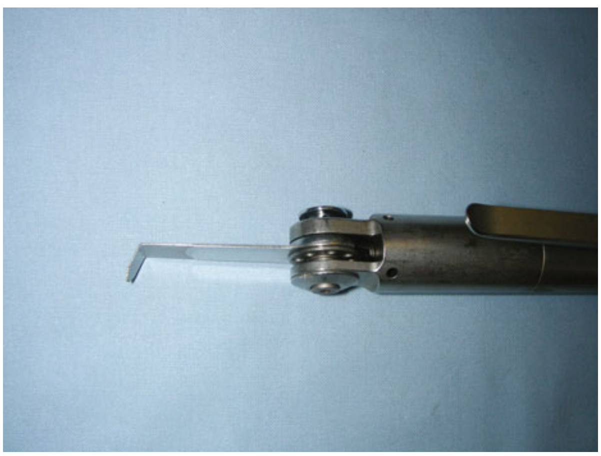

Fig. 16. An oscillating saw with an angled blade to enable sternum osteotomy from a distantly set skin incision

7.2.3 Discussion

The until now established technique of Ravitch (Chapter 7.1) includes determined standards, independent of the size or the localization of the keel chest deformity. The pectoralis major and rectus abdominis muscles are elevated as muscle flaps from the sternum and the parasternal rib cartilages. They are reflected to lateral as well as to caudal to expose disfigured rib parts. Such a creation of extended wound areas in the submuscular area with extensive desinsertion of musculature does not appear absolutely necessary for the intervention at the skeletal structures. Therefore, a modified access utilizing limited transmuscular approaches on every individual levels of the disfigured single rib seemed to be a practicable alternative. Through solely blunt separation of muscle fibers, it hardly comes to hemorrhage and therewith blood loss is negligible. It furthermore appears as a special benefit that such blunt muscle preparation and the avoidance of muscle flap elevation lead to substantially lesser consumption of postoperatively required analgesics [25]. On the one hand, this is because the invasive wound surfaces remain far smaller and almost all of the muscle fibers remain inserted. A peridural catheter which is usually proposed in the standard techniques can therefore be left off except for cases with very extensive deformities. As a further consequence of such diminished pain phases probability of earliest discharge from the inpatient treatment ensues.
The particular advantages in performing the muscle split technique consist of:

- substantially reduced wound area,
- less hemorrhage,
- remaining muscle and fascia insertions,
- almost no risk of muscle retraction,
- immediate postoperative mobilization, and
- unimpaired postoperative range of shoulder motion.

Minor disadvantages consist of:

- lesser overview to rib cartilages and
- more risk for minimal pleura lesion.

If a sternumosteotomy is not necessary and only parasternal chondrotomies are applied, however using

the muscle split technique, full mobilization of the shoulder belt already after few days without strain is permitted. These exercises can be upgraded then after the third postoperative week, which is in explicit contrast to the conventional technique, where several more weeks more of rest to relieve the muscle sutures is advised, particularly in adolescents and adults. For the case that sternum osteotomy is carried out simultaneously within the remodeling procedure, the full exercise strain is permitted first after the sixth postoperative week whereby however from the third postoperative week already, based on the remaining muscle insertions, full passive mobility is permitted.
The use of absorbable osteosynthetic material (Figs. 9 and 10) additively supports bone healing, whereby this means no essential advantage versus the utilization of wire cerclages. Nevertheless wire cerclages are permanently remaining foreign objects and can cause local problems such as skin irritation or perforation by loosening after months or years. Occasionally these wire cerclages are permanently palbable or even well visible in slim patients. On the contrary, absorbable osteosynthetic material is resorbed by autolysis (Fig. 17) within 1–2 years completely [2, 16, 17, 24, 25]. Additionally, these devices can be cut with scissors or hot electric loops and tailored according to the size required. Manipulation or bending at room temperature is possible without affecting their performance. Thus, they can be formed exactly enabling reliable and secure bone and cartilage fixation to allow precise sternal reposition. These plates and screws give the surgeons unlimited possibilities to reshape and to adapt the bone-cartilage complex to the defect with minimal manipulation and injury of blood supply.

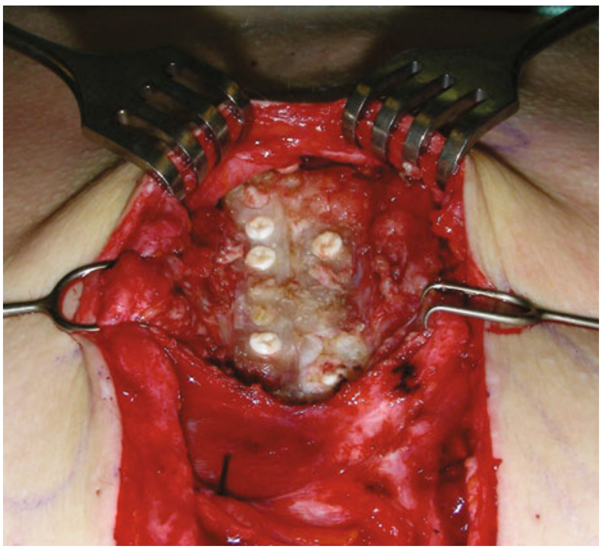

Fig. 17. Absorbable osteosynthetic material during the process of disintegration in a case with minor corrective surgery of remnant rib humps 1 year after remodeling surgery

Fonkalsrud in 2004 [7] described a modification of the Ravitch technique using temporarily implanted metal struts for 6 months. The argumentation was that such a metal strut produces stability to the anterior chest wall during the healing phase and therewith prevents the danger of recidive development with sternum protrusion. Abramson recently described a modification of the Nuss procedure but in contrast to that using the pectus bar as a compressive device, placed subcutaneously or beneath the pectoralis muscles. The results seem to be promising so far, avoiding extensive surgery and median scars [2]. However an essential disadvantage is that this strut must be removed after 6–12 months and therewith further surgery and hospitalization is necessary. Absorbable plates and screws for sternum refixation and a postoperative keel chest brace circumvent this necessity of a rigid internal fixation and a second intervention. Already Matsui and Gürkök applied large bioabsorbable copolymer plates and screws for the correction of pectus excavatum deformities, but not in the carinatum deformity [9, 16]. However, such voluminous and thick absorbable plates are not desired because they are very well palpable and visible through the presternal skin and furthermore yet require a substantially long absorption time. The use of small platelets and screws (Fig. 9) in connection with postoperative bracing permits a similarly rigid immobilization of the anterior thoracic wall for the period of bone healing and scarring of the reefed perichondrium sleeves. The use of such absorbable osteosynthetic material (Bionx™, Tampere, Finland) for pectus carinatum surgery until now caused no complications at all [24, 25].
Moreover, an additional protection through immobilization of the osteotomized and refixed sternums is produced using the keel chest brace postoperatively (Chapter 7.4), what yet moreover eases the postoperative mobilization maneuvers. Bearing the keel chest brace permanently for 23 h daily contains a further advantage, namely the permanent compression of the scar over a period of 2 months. This causes, also well known from the literature [29], a reduction in the probability of development of inadvertent scar formation (Chapter 12.2). The distinct compression effect of the brace paddle to the scar is

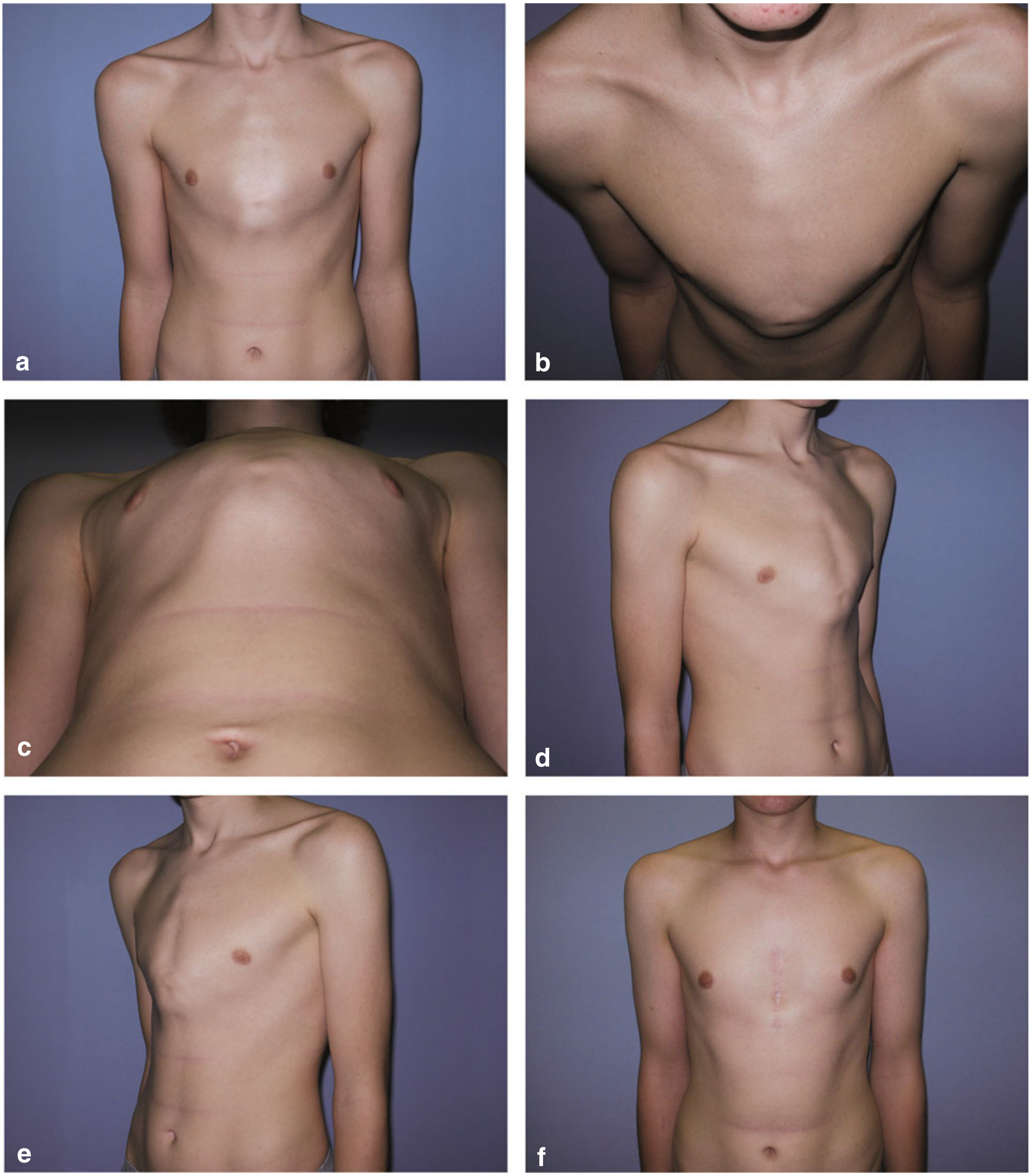

Fig. 18a–e. Preoperative situation in a 16-year-old male with symmetric chondromanubrial deformity. **f–j** Situation 1 year postoperatively after modified repair with muscle split technique and 2 months of keel chest bracing.

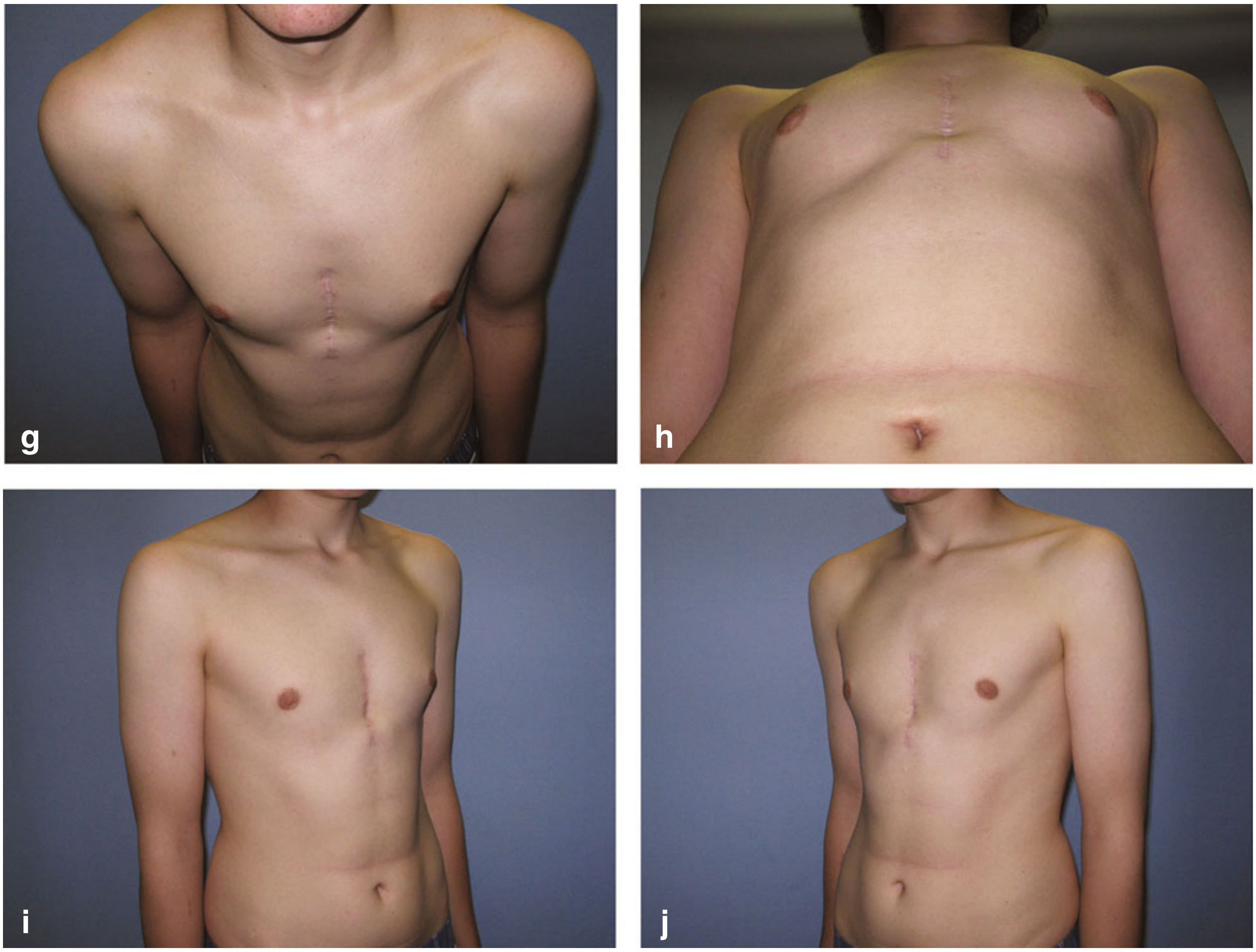

Fig. 18. (*Continued*)

evidently visible in Fig. 7 of Chapter 5.1 as well as in Fig. 4 of Chapter 7.4. A silicone containing surface mounted on the compression paddle additively successfully avoids hypertrophic scar development (Figs. 18 and 19).

Nevertheless a slight disadvantage must be noted such as the transmuscular access yields a somewhat limited view to the rib cartilages and perichondrium. Particular caution must be deployed because of the relatively narrow transmuscular access, especially in individuals with well-trained bulky muscles to avoid injuries of pleura and lung. If the deformity is extending very laterally, the muscle separation must be carried out very carefully and ideally with the aid of magnifying loupes to protect the pectoral nerves hereby passing partially oblique to the muscle fiber direction. Even partial or minor nerve branches severing causes partial muscle atrophy and diminishes function and aesthetic appearance.

For that reason and in marked deformities, the conventional approach with muscle flap elevation for remodeling still is suggested to circumvent nerve lesions. Anyhow the muscle split technique can be applied in the caudal thoracic area at the rectus abdominis muscle also (Fig. 20).

Comprehensively we recommend the use of the muscle split technique in patients suffering with lesser or moderate Pectus carinatum deformity, if not more than five rib cartilages must be incised or shortened. In cases requiring access to more than five ribs on each side the conventional Ravitch procedure is still the first choice of approach. However, no evidence-based studies comparing the Ravitch procedure with the modified muscle split technique exist to date. A

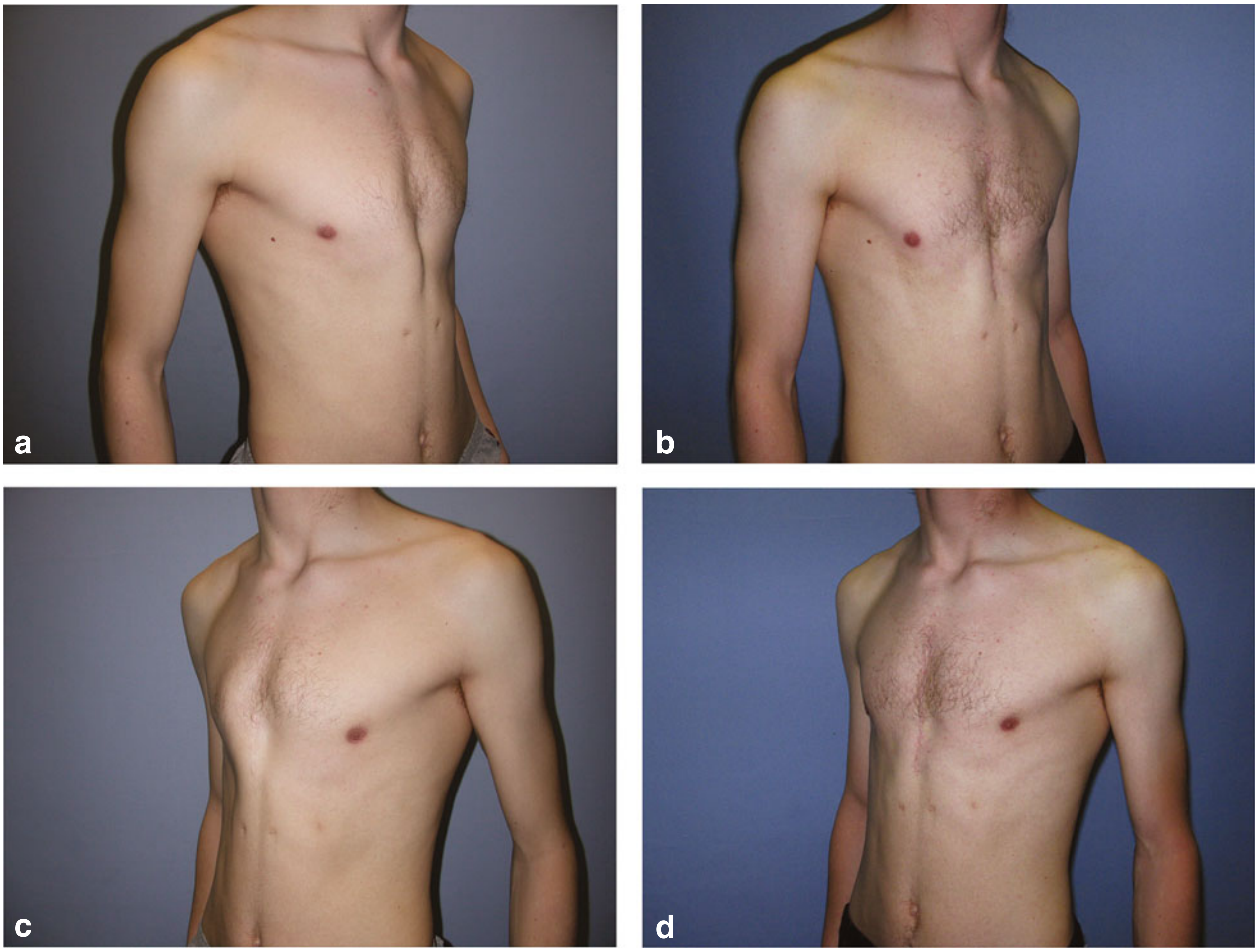

Fig. 19a, b. Preoperative situation in a 19-year-old male with moderately asymmetric chondromanubrial deformity. **c, d** Situation 1 year postoperatively after modified repair with muscle split technique and 2 months of keel chest bracing

prospective study design seems no longer be possible because based on previous and ongoing experiences it now is considered essentially to avoid pains and long-term periods of immobilization. Therefore the conventional technique in the mild and moderate deformities has been abandoned at our institution for the benefit of the patient.

Solely compressive orthotics [10, 13, 27] without surgery may diminish the extent of minor deformities if applied early in childhood and if applied for many months, depending on a long-lasting endurance of the affected patients. However, to our experience only the combined treatment of surgery with postoperative compression provides with the best aesthetic outcome so far but resulting in a surgically produced scar.

7.2.4 Conclusion

Our previous experiences showed that the modification of the conventional Ravitch technique, according to the Innsbruck concept with muscle split technique, use of absorbable osteosynthetic material as well as the postoperative keel chest brace, reduce the entire morbidity of the intervention itself. Thus the postoperative rehabilitation phase for the patients is substantially shorter and remains more comfortable. Consistently good to perfect aesthetic results after keel chest surgery however may differ in using these different techniques, namely both the conventional Ravitch procedure or the modified Innsbruck approach. This may not be ascribed to the muscle split technique alone but rather to the integrated treatment with utilization

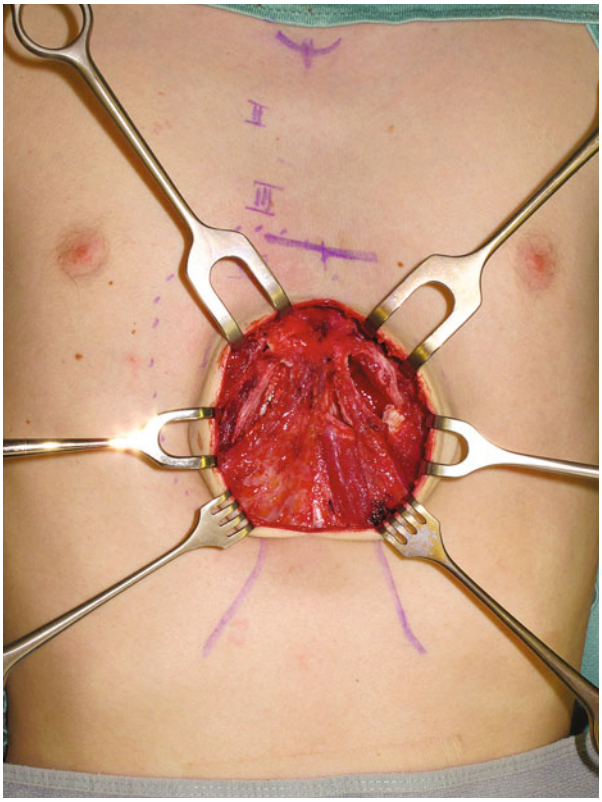

Fig. 20. Longitudinal muscle splitting at the rectus abdominis muscle in order to expose and enable partial excision or transection of the lowermost rib cartilages

of a keel chest brace, which on the one hand permits a more stable support for healing of the severed skeletal structures and on the other hand minimizes the risk for the development of hypertrophic scars or even keloids.

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7.3 Cartilage chips for refinement after keel chest remodeling

Anton H. Schwabegger

This chapter, in analogy to Chapter 6.6.1, describes the appropriate use of cartilage chips in the corrective refinement of remaining depressed deformities after or during the surgery of asymmetric or mixed (pectus arcuatum) expressions of keel chest deformities. The detailed technique is described in the referred chapter and substantially does not differ in harvesting, comminution, and wrapping into surgicel® as well as reimplantation into subcutaneous pockets.

Usually plenty of excised rib cartilages is availabe after the standard Ravitch procedure in the keel chest surgery, commonly subject of biological disposal. Instead of wasting these abundant cartilages, they all or in parts may be utilized, whenever appropriate, to fill up concomitant contour defects to some extent.

Such refinements by utilization of autologous cartilage grafts can be required, when irregularities in the contour of skin surface of the anterior thoracic wall e.g. develop

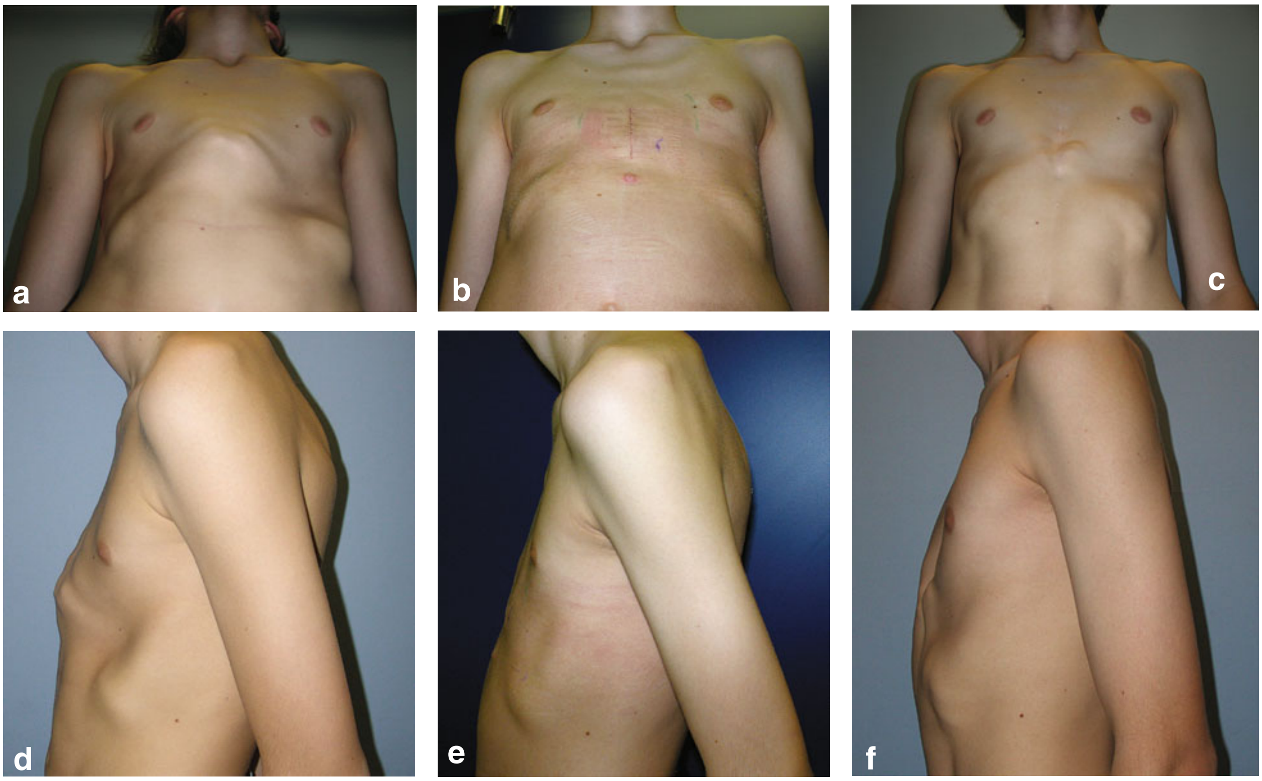

Fig. 1a and d. 16-Year-old male patient with almost symmetric keel chest deformity at the sternum area but and predominately left-sided depression of the lower anterior thoracic wall, rather unapproachable for conventional remodeling surgery. **b** and **e** Situation 8 weeks postoperatively, modified Ravitch procedure with muscle split technique (Chapter 7.2), after removal of the keel chest brace. The left-sided depression was filled up with cartilage chips resected from abundant parasternal cartilage parts. Notice the achieved and very satisfactory resulting symmetry of the lower thoracic wall. **c** and **f** Long-term follow-up 12 months after surgery with partial resorption of transplanted cartilage chip grafts but still with satisfactory result. Unfortunately some minor rib hump developed at the left paraxiphoid area, whether as a genuine relapse or eventually also caused by dislocated cartilage chips

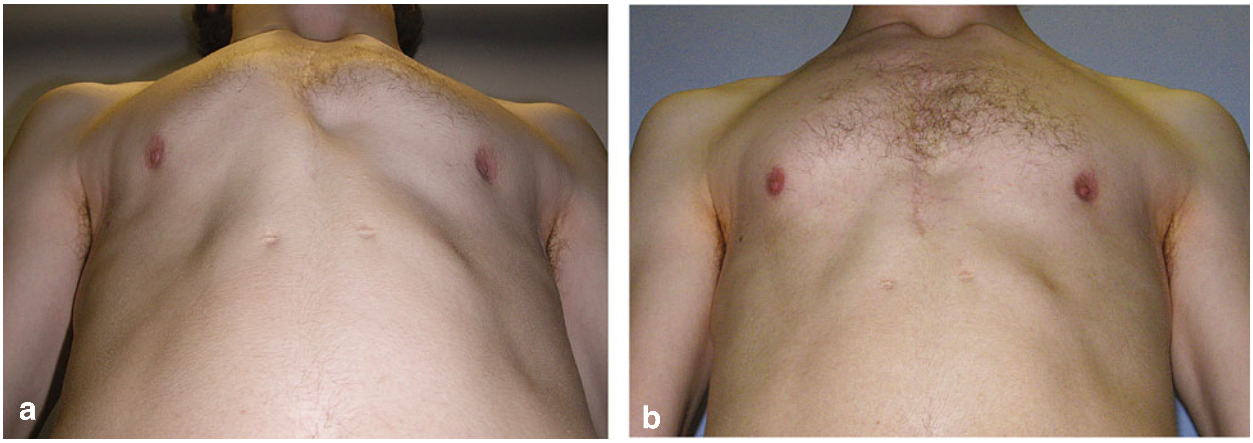

Fig. 2a. 18-Year-old boy with asymmetric keel chest deformity and a central depression between the keel ridges, additional depression at the left infrapectoral area. b Same patient 12 months after keel chest correction according to the Ravitch procedure, modified with muscle split technique, simultaneously then the central depression and the infrapectoral areas were sucessfully filled up with subcutaneously placed cartilage chip grafts, resulting in a smooth and rather symmetric skin surface

after keel chest surgery, during further growth of the body until adolescence. However, such refinements may also be necessary during keel surgery itself to augment already preexisting depressions of the thoracic wall adjacent to the keel deformity. Such accompanying depressions, if present at all, are mostly located at the lateral border of the keel-humps or extend along the inferior rib arches (Fig. 1a and d). Occasionally depressions may also be found at the central sternal region, dividing the keel into two ridges (Fig. 2a). Other depressions are not accessible or manageable by the keel remodeling surgery itself. They may present themselves rather distant from the site of surgical access, spread very laterally thus remain inaccessible for yet justifiable surgery (Fig. 3), or the convexity simply is too largely scaled to allow further invasive rib remodeling.

Furthermore, this technique of autologous cartilage chip transplantation may also be extraordinarily helpful after corrective surgery, when partial relapse of keel deformity or isolated rib hump formation occurs and depressed areas are present simultaneously. Every autologous non vascularized tissue graft and also cartilage chips may undergo unpredictable resorption or alteration of volume due to minor blood supply, infection, or dislocation, which must be part of the informed consent for those patients treated with that additional refinement.

Fig. 3. 12-Year-old female with asymmetric keel deformity and major depression at the left inframammary region. This arcuation of the lower ribs will not easily be manageable by rib remodeling procedures but may rather be filled up with autologous cartilage grafts, harvested from the left parasternal deformed rib parts. Patient is scheduled for surgery at the age of 16 or even no surgical intervention may be necessary, depending on the further development of the breasts

References

See Chapter 6.6.1.

7.4 Special after-treatment, the keel chest device

Barbara Del Frari, Anton H. Schwabegger

Compressive orthotics in former times were used for the treatment of pectus carinatum, but usually failed to therapeutic success. This was due to the fact that overgrowth of rib cartilages may be bent flat temporarily by application of continuous compression lasting for several months [4, 6]. Usually the overgrowth of rib cartilages in length causing the deformity may not be treatable just by compression, especially in adolescents and adults. On the other hand, the experiences of orthopedics in the construction of such orthotic compressive chest braces are available to apply such devices just in the postoperative keel chest treatment. Such an adjunctive brace should serve for immobilization and lowering the respiratory excursions of the chest wall and sternum during the healing process. That kind of brace changed its task from a primary treatment device to a secondary supportive treatment tool to maintain the surgically achieved shape and to enable early healing of mobilized sternum bone parts and rib cartilages. Furthermore, due to restriction of excessive inspiratory excursions it avoids rupture of deeply set sutures, such as perichondral reefing or muscle sutures. Postoperatively after keel chest correction with the conventional Ravitch procedure (Chapter 7.1) or the modified Innsbruck protocol (Chapter 7.2) we routinely first apply a circumferential elastic compression dressing (Fig. 1) with an adhesive under-surface for the fixation and compression of a steel wool padding above the area of remodeling to immobilize the chest wall. At about the third postoperative day, all patients receive an individually manufactured "custom-made keel chest brace" with moderate compression that is applied in a sagittal direction (Fig. 2a–c). The compression brace has to be worn for at least 23 h/day for about 6–8 weeks to guarantee healing continuous immobilization and compression of the shaped area of the thoracic wall [7, 8]. The purpose of the brace is to provide further stability additionally to perichondral reefing sutures and, occasionally applied, bioabsorbable osteosynthesis. The brace eliminates postoperative respiration movements at the thorax, which furthermore reduces pain, but still permits deep respiratory excursions during breathing therapy postoperatively and later on after demission physical exercise by abdominal breathing using the diaphragm.

A silicone containing foamy sheet (Fig. 3a, b) is attached to the anterior pad encompassing the area of remodeling and the presternal skin scar to prevent the development of a hypertrophic scar (Chapter 12.2). So far in our series of surgical keel chest with ensuing bracing hypertrophic scarring was apparent in no case. In two particular cases even with precedent open-heart surgery subsequently a hypertrophic scar developed and remained as a clearly visible stain. Years thereafter keel chest surgery was performed using the same access, however with excision of the whole length of the scar for intentional scar correction. The resulting scar after keel chest correction and ensuing brace compression therapy for 2 months led to a much better appearance than the remaining, also excised and equally surgically treated scar at the sternum, however lying beyond the reach of silicone pad com-

Fig. 1. Circumferential elastic compression with self-adhesive dressing, steel wool bolster beneath the area of remodeling to provide with smooth compression

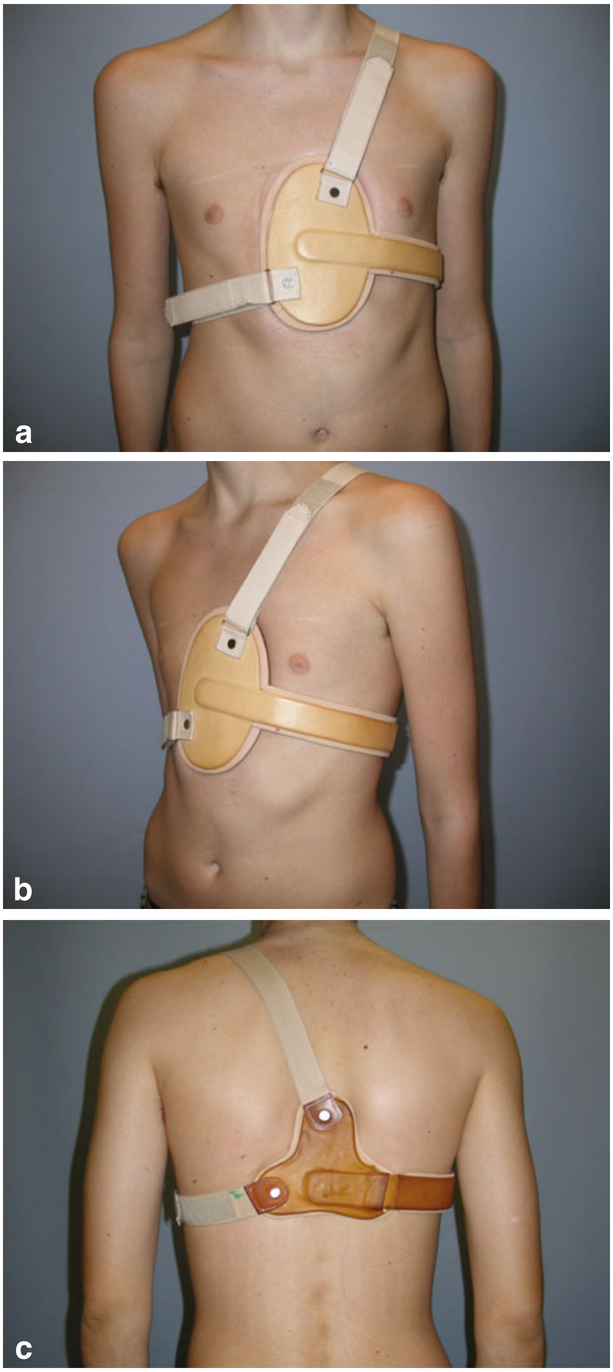

Fig. 2a. Anterior aspect of a custom-made keel chest brace well fitting and covering the area of surgical remodeling of the affected skeletal structures. Notice the metal wing at the left side and the adjustable flexible band at the right side. **b** Lateral aspect of the same brace, the metal wing smoothly adapted (custom-made) to the patient's individual chest shape. **c** Dorsal aspect depicting the counter-part of pad for harmonic balance of the pressure applied in a sagittal direction and protection of the skin against undue compression along the dorsal spinal processes

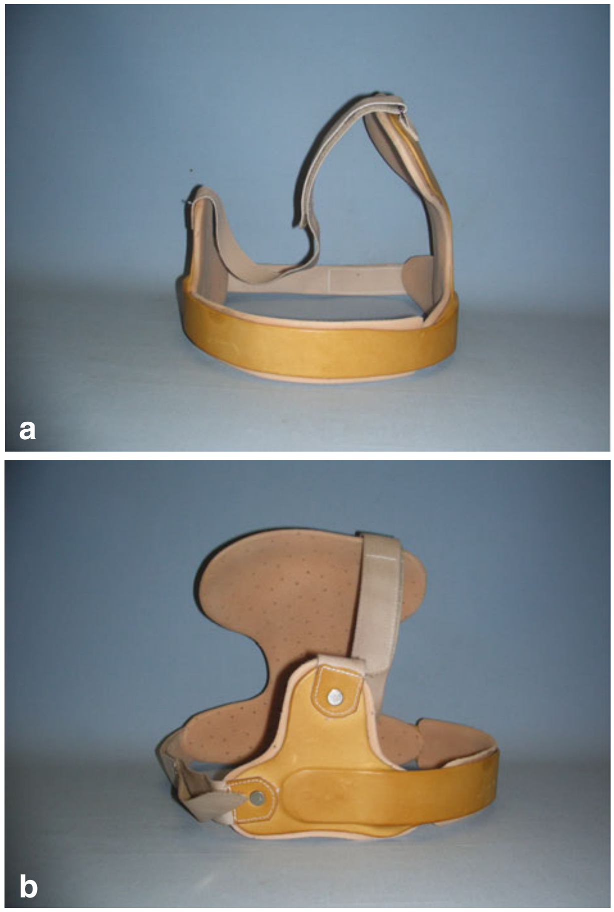

Fig. 3a. Lateral (right) aspect of a custom-made keel chest brace ready for use, showing a steel wing (front) covered with skin compatible leather surface. **b** Dorsal aspect to depict the anterior compressive pad for maintaining the shape of thoracic wall achieved through surgery, which furthermore is covered with a silicone sheet to reduce scar formation and above that to prevent hypertrophic scarring

pression (Fig. 4). One might argue that the different ages of both patients at heart and then at keel chest surgery could have been responsible for the different developments of the appearance of these scars, but on the other hand the curative effect of compression on hypertrophic scars is well known and among others [1, 9] an established way of therapy [2, 3, 5]. As clearly visible in Fig. 4, the exactly circumscribed flat and pale segment within a very long though fresh looking and unsightly scar may evidently be attributed only to the beneficial effect of the compression pad that exactly covered this now pleasantly looking part of the scar.

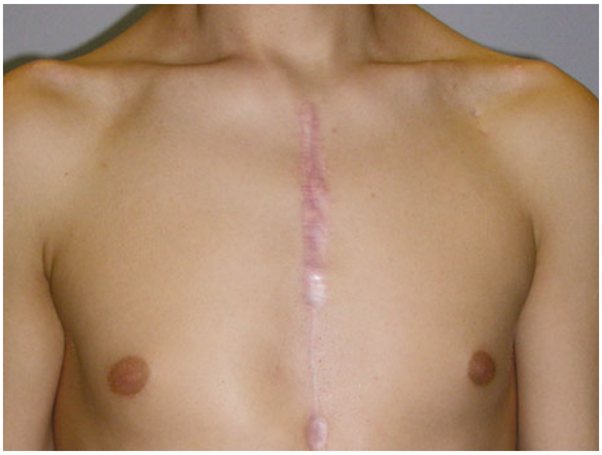

Fig. 4. Keel chest surgery and simultaneous scar correction performed many years after heart surgery at childhood. Notice the clear demarcation of the area which underwent compression by the pad of the keel chest brace (lower part of scar with pale surface) versus the sections which remained free of pressure remote from the pad (xiphoid and cranial part of sternum)

In our series of more than 60 patients who underwent keel chest surgery since the utilization of the keel chest brace no hypertrophic scarring was apparent within a range of follow-up from 6 weeks to 36 months.

Especially in females and herein in asymmetric cases with surgical access along the inframammary crease, specially designed braces (Figs. 5 and 6) can be manufactured for adequate inframammary compression, extending up into the décolleté.

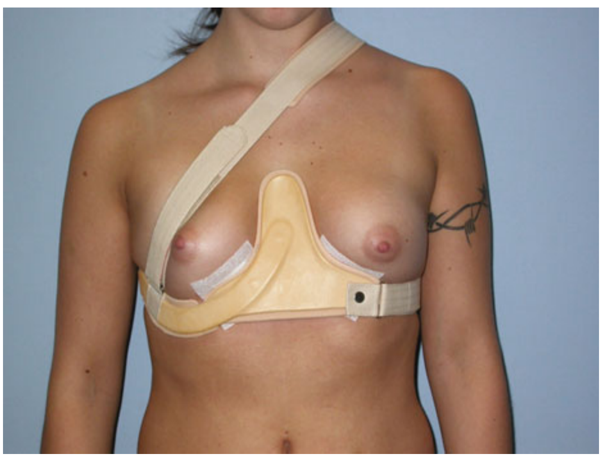

Fig. 5. Specially designed brace for a female who had bilateral incisions for keel chest surgery at the parasternal inframammary crease including horizontal sternum osteotomy

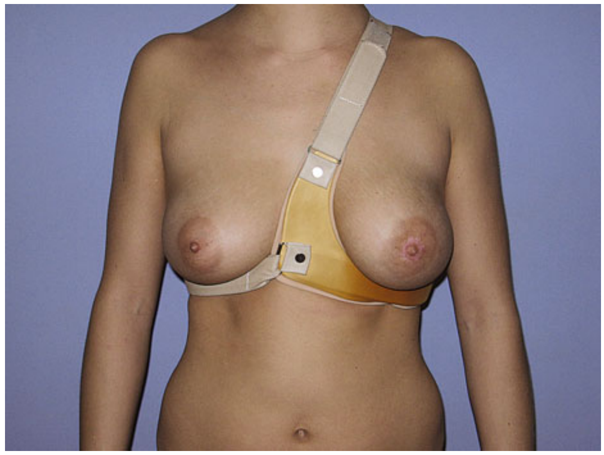

Fig. 6. Particular design for an adult female, who underwent unilateral chondrotomies through a single inframammary incision on the left side. The compressive pad reaches also up to the décolleté region, where chondrotomies were performed from a subcutaneous and transmuscular approach through that inframammary incision

There were minor postoperative complications such as epidermal skin irritations and formation of blisters caused by inappropriate initial pressure from the keel brace in singular cases (Chapter 12.1). In one of our cases unfortunately 1 week postoperatively and 2 days after explantation of the peridural catheter the patient developed meningitis, caused by skin bacteria. The elastic compression dressing, obligatory in every keel chest patient immediately applied after wound closure, was accused to eventually have deteriorated the detection of local skin infection with ensuing penetration of the infection into the dural surfaces. That assumption could not be verified, however every location (skin perforation) of peridural catheter implantation at the back should be excluded from compressive dressings for early detectability of any irritation.

The custom-made keel chest brace usually is manufactured and available within 2–3 days after measurements, which may take place at the second to third postoperative day. The patients are dismissed not before the brace fits well, then are obliged to sleep on the back for 6–8 weeks postoperatively, as long as wearing of the brace is necessary. The period of application depends primarily on the extent of the pre-existing keel deformity, the extent of surgery, and the overall morphology of the patient. Athletic tall patients with excessive deformity may even require longer periods of bracing, whereas in unilateral deformities in youths 4 week may be sufficient. However, taking into consideration the potential development of hypertrophic scars it may be wise in every case to suggest bracing for 6 weeks at least.

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Special techniques in pectus arcuatum and mixed deformities

8

Anton H. Schwabegger

The pectus arcuatum deformity [1, 3] with minor or major curvature of the sternum, including either symmetric excavatum and carinatum features along a longitudinal (Figs. 1 and 2) axis or asymmetrically along a transversal axis (Figs. 3 and 4) needs a comprehensive approach.

Surgical repair of minor arcuatum deformities (Figs. 1 and 2) may be corrected using the conventional or

Fig. 1a and b. Boy of 13 years of age with symmetrical pectus arcuatum deformity, without cardiopulmonal impairment. **c** Intraoperative situs, bioabsorbable plates in situ to fix the newly shaped sternum, which was twice osteotomized horizontally to unbend the curvature to an almost frontal plane. The remaining excavatum deformity is filled with cartilage chips derived from the resected ribs. **d** and **e** Same patient 4 years after surgery and keel chest bracing for 6 weeks

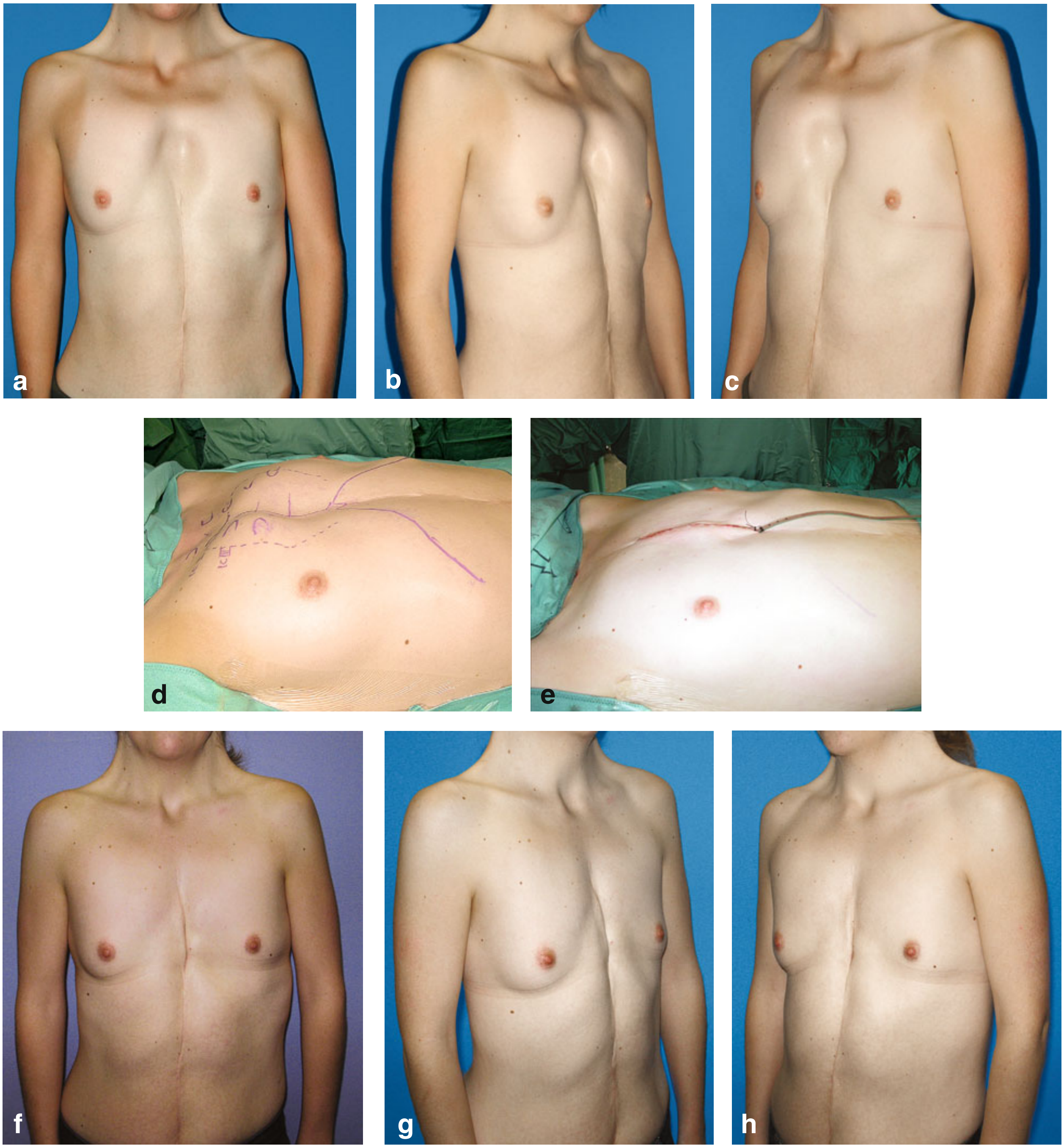


Fig. 2a–d. 20-Year-old female with characteristics of Noonan syndrome, webbed neck, minor body height, and pectus arcuatum deformity. She underwent omphaloschisis surgery at early childhood. **e** Intraoperative situs with planned access for osteochondroplastic remodeling, including the utilization of resected cartilage chips for refinement. **f–h** Same patient 35 months after remodeling and postoperative bracing. Beneficial scar correction was refused by the patient

modified Ravitch technique with chondrosternoplasty (Chapters 6.1, 6.2, 7.1 and 7.2). Resected bone and cartilage wedges may be reimplanted at sites with osteochondrotomies to fill up distending gaps (Fig. 5). Furthermore, resected abundant cartilages can be gently minced and used as autologous cartilage chips to fill up minor remaining deformities (Fig. 1c) otherwise hardly accessible without major interventions (Chapters 6.6.1 and 7.3).

Extensive pectus arcuatum or pouter pigeon breast [2] deformities along a longitudinal axis, either symmetrically or with minor asymmetry are repaired following the principles of chondrosternoplasty after Ravitch, individually containing minor or major modifications (Fig. 3), which depends on the shape of deformity and age of the patient. Especially in the pouter pigeon breast with premature ossification at the angle of Louis, a wide transverse bony wedge resection is necessary to enable sufficient repositioning of the curved sternum (Fig. 5), preferably followed by stable osteosynthesis either with bioabsorbable mesh plates and support of the elevated lower part of the sternum with alloplastic material or even with metal struts to achieve a permanent shape [5–7].

The repair of predominantly asymmetrically shaped pectus arcuatum or carinatum deformities may be quite simple or extensive (Fig. 3) on the other hand, depending on the extent and shape of the deformity at each side. Postoperative compression of the reshaped anterior thoracic wall with a custom-made keel chest brace

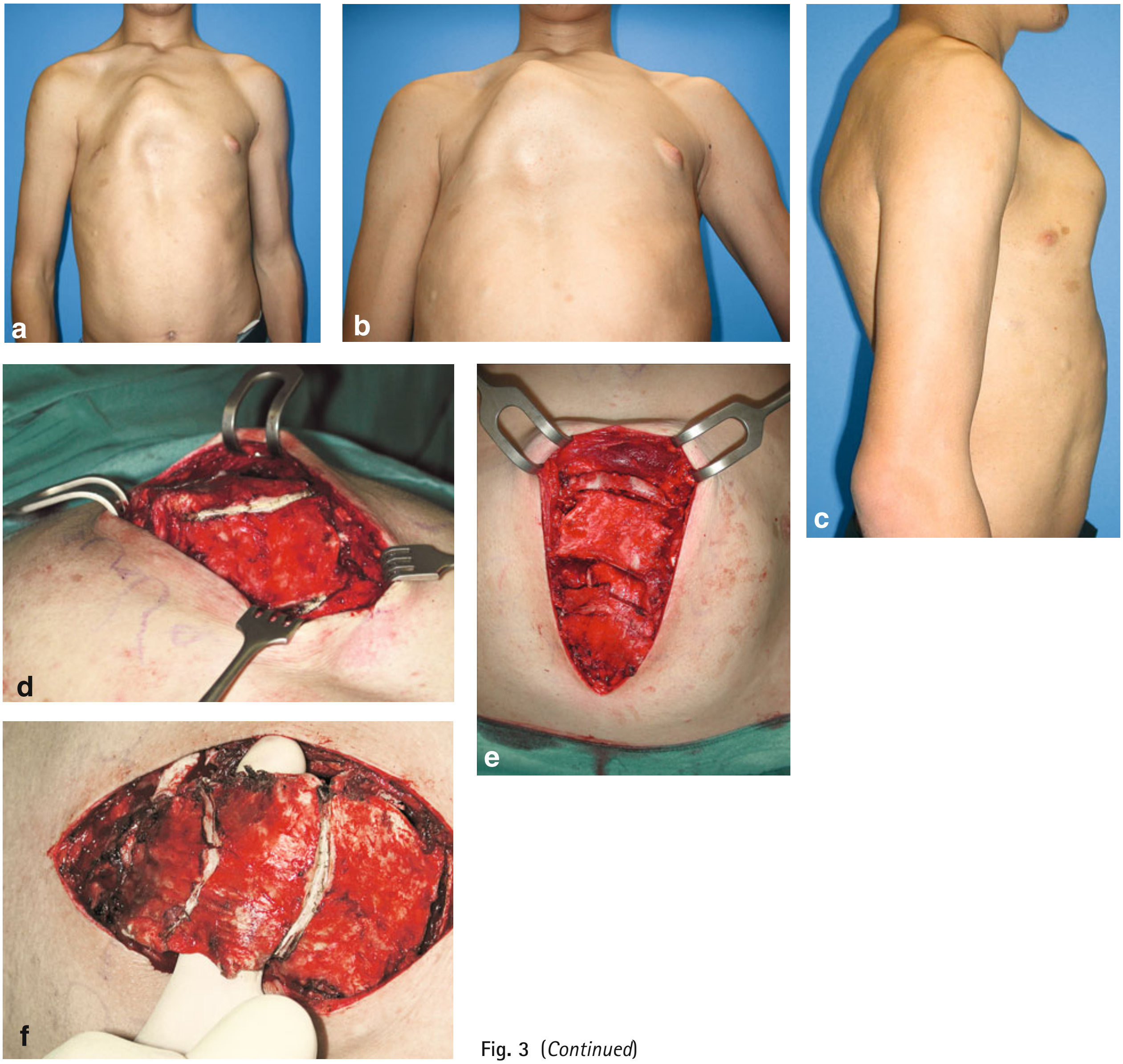


Fig. 3 (*Continued*)

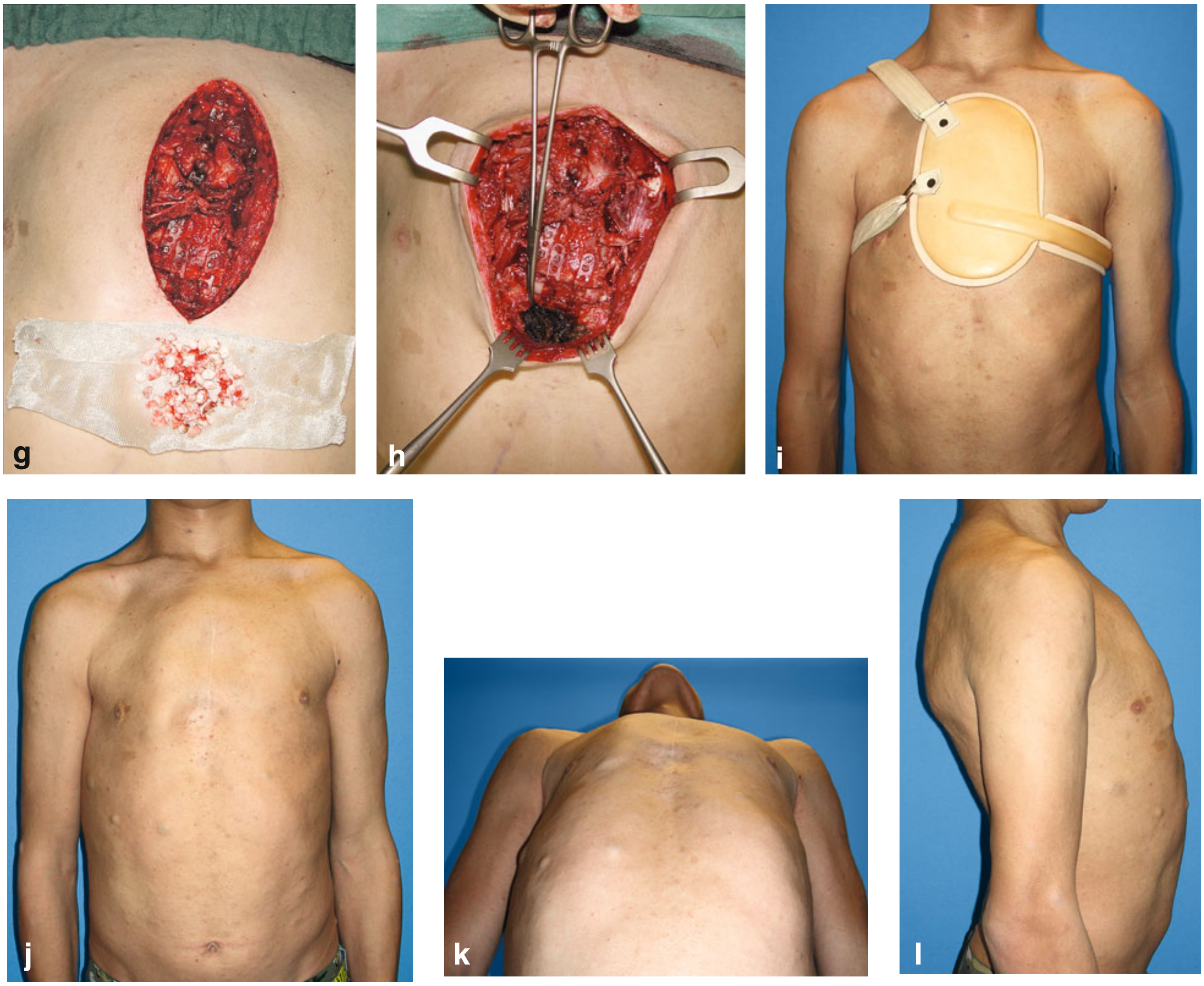
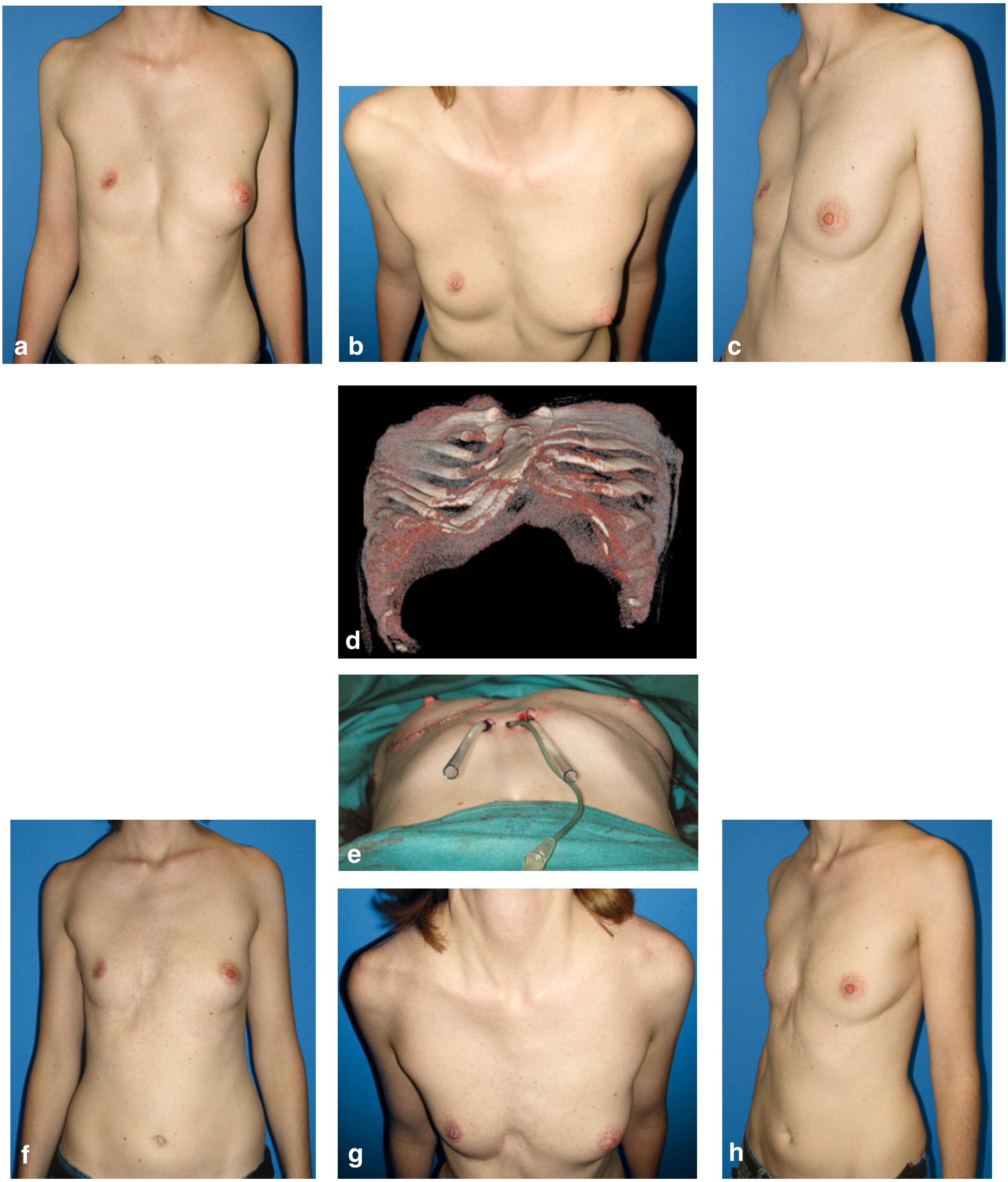


Fig. 3a–c. 18-Year-old very tall male with extensive pectus arcuatum deformity, predominantly carinatum features, associated with Recklinghausens disease. **d–h** Intraoperative situs with exposed projecting broad sternum, periosteum incised (**d**) prior to multiple horizontal osteotomies and bony wedge resections (**e**). Intrathoracal structures bluntly mobilized from posterior surface of the sternum (**f**). After multiple osteotomies and shaping of bone remnants, a small wedge of bone is reimplanted cranially. The refixation of bones is administered with bioabsorbable plates and screws (**g**). Resected cartilage (**h**) is used to fill up a minor depression at the caudal area of the remodeling. **i** Keel chest brace was worn for 2 months to temporarily stabilize the area of remodeling until healing of the mobilized skeletal structures. **j–l** Same patient 4 years after a single access corrective surgery of major pectus arcuatum deformity

Fig. 4a–c. 29-Year-old woman with severe unilateral mixed thoracic deformity, predominantly hemithoracal hypoplasia without signs of Poland sndrome. **d** Volume rendering 3D scan of the same patient revealing major malrotation of the sternum to the right and multiply narrowed intercostal spaces at the right hypoplastic thorax. **e** Same patient immediately after completion of combined surgery with minor open access, implantation of one pectus bar, sternum derotation, and multiple parasternal chondrotomies and chondrectomies with utilization of cartilage chips to augment the space behind the right breast, the cartilages implanted behind the Pectoralis major muscle. **f–h** Same patient 1 month after explantation of the pectus bar, which remained in situ for 16 months. The shape markedly improved but still insufficiently symmetric


a
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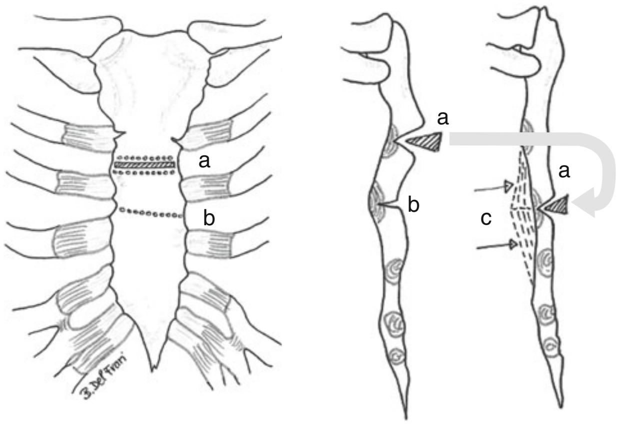

Fig. 5. Scheme shows hypothetically placed horizontal bony incisions (*a*, *b*), the site at the intercostal levels depend on the shape of the deformity. A bony wedge resected at the prominence (*a*) may be placed at the raw exposed incision site at the area, where the sternum is bent forward (*c*) and thus elevated. This wedge will be fixed with osteosynthetic material to maintain its shape until bony healing is completed

(Fig. 3e) is mandatory to allow healing with a permanently stable outcome. Especially in cases with sternum malrotation the surgical correction may develop to an endeavor, if symmetry should be achieved (Fig. 4). In such cases usually underdevelopment of the whole hemithorax complicates any attempt for volume augmentation. Pectus bars may not be able to sufficiently support an elevated sternum due to sunken funnel rims and ribs at the affected side with no counter bearing options for it. In such cases only limited options exist for remodeling, combining methods like pectus bar with partially open remodeling, derotation of the sternum by osteotomy, and tissue augmentation by lipofilling or silicone implants. Cartilage chips, if available (Fig. 1c) may complement minor reshaping attempts. Even invasive microvascular sternum turn-over [4] might be taken into consideration, however also performed as a combined approach with metal struts for reinforcement of the temporarily destabilized thoracic wall.

8.1 Conclusion

For a propitious remodeling of pectus arcuatum and other mixed deformities, the whole armamentarium of remodeling options including autologous tissue reimplantation may be available. Favorably postoperative treatment with compressive devices like the keel chest brace will enhance the aesthetic long-term outcome with respect to the shape itself but also to the scar at the critical region in the décolleté (Chapters 7.4 and 12.2).

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Special aspects in females

Anton H. Schwabegger

The minimally invasive correction of the funnel chest deformity utilizing metal struts [1, 9, 10] refined by Weber and Huemmer to the new Erlangen technique [25] as well as by Nuss [14] to the so-called MIRPE (Minimally Invasive Reconstruction of Pectus Excavatum) technique nowadays predominantly is carried out in children and adolescents [5]. This type of surgical correction at this age, if indicated by medical reasons, is ideal because the thoracic skeleton in particular the anterior thoracic wall is present with yet sufficient flexibility in order to give way to the pressure by a transsternally implanted metal strut or retrosternally placed pectus bar. The original MIRPE method on the other hand however is the fewer an ideal treatment method, the older patients are. Because with body maturation at adulthood the skeletal structures increase their rigidity thus by methods of internal suspension, likewise with a metal strut, the thoracic wall becomes less moldable [7, 26]. However, many patients seek the contact of the physician for the first time in adulthood with a strong desire to have their deformity at the breast corrected. With minor or only moderate extent of pectus excavatum deformities and absence of cardiopulmonal restriction silicone implants may very well fulfil the requirements of presternal volume augmentation, as well as lipofilling [3, 18] may do that (Chapters 6.5 and 6.6.2).

On the other hand however, when extensive deformities exist, occasionally also with slight or moderate cardiopulmonal restrictions, the remodeling of the anterior thoracic wall utilizing adequate methods or techniques seems to be justified [17, 20]. Fonkalsrud in 2004

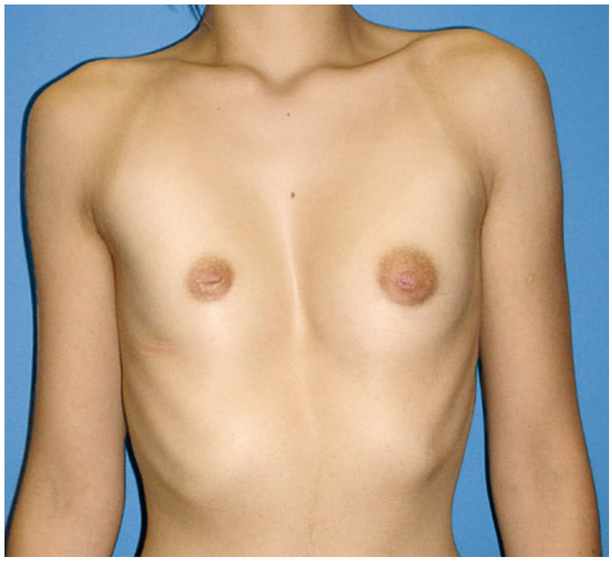


Fig. 1. Strabism of the breasts caused by an underlying asymmetric pectus excavatum deformity with sternum malrotation and conjoined minor breast volume asymmetry. The latter is evidently aggravated by the underlying skeletal asymmetry

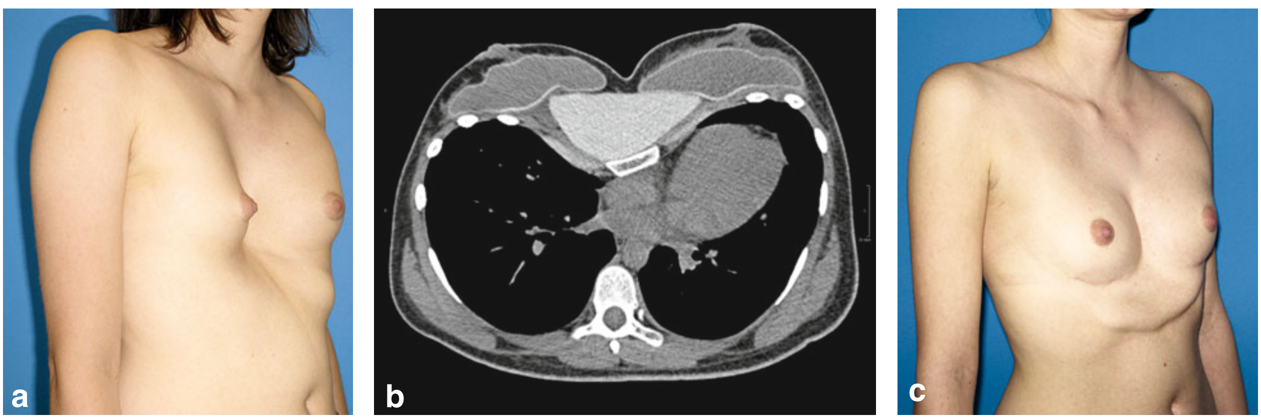


Fig. 2a. Severe pectus excavatum deformity in a 20-year-old female. **b** Situation seen at CT after inset of three implants, one centrally to fill up the pectus cavity and two breast implants. **c** Situation after implantation of a further custom-made silicone implant horizontally at the epigastric region with gravitational effect and dislocation resulting in a well visible subcutaneous alien

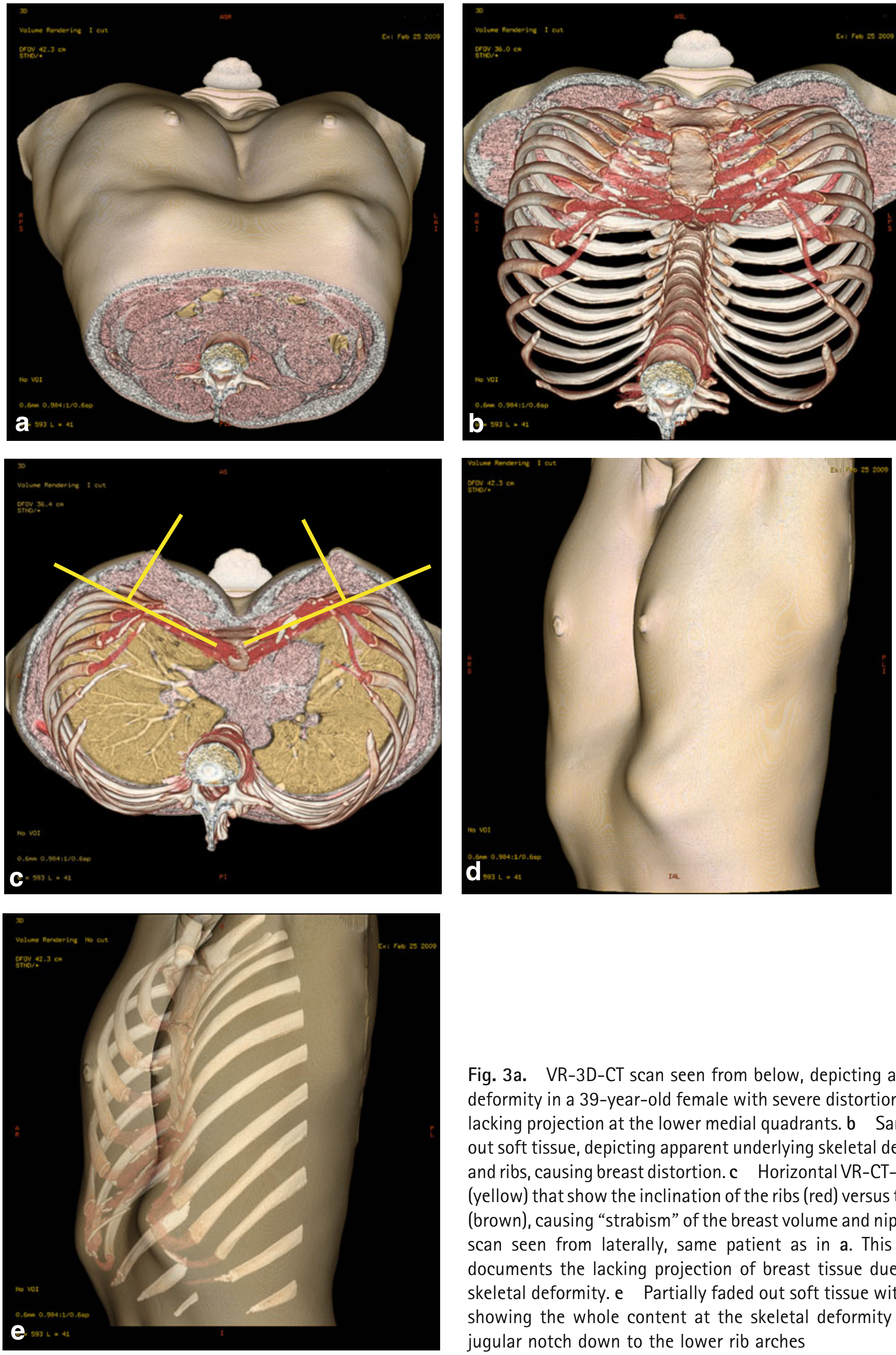


Fig. 3a. VR-3D-CT scan seen from below, depicting a pectus excavatum deformity in a 39-year-old female with severe distortion of the breasts and lacking projection at the lower medial quadrants. **b** Same view but faded-out soft tissue, depicting apparent underlying skeletal deformity at sternum and ribs, causing breast distortion. **c** Horizontal VR-CT-scan with markings (yellow) that show the inclination of the ribs (red) versus the sunken sternum (brown), causing "strabism" of the breast volume and nipples. **d** VR-3D-CT scan seen from laterally, same patient as in **a**. This kind of view also documents the lacking projection of breast tissue due to the underlying skeletal deformity. **e** Partially faded out soft tissue with transparent view showing the whole content at the skeletal deformity reaching from the jugular notch down to the lower rib arches

reported that out of 104 females with pectus excavatum, nine had successful pregnancies prior to deformity repair, but all suffered from third trimester dyspnea [8]. Under these aspects, the repair of funnel chest in females gains considerable medical attention.

Above all in girls and women, the unconcealable stain of a funnel chest deformity leads to considerable timidity, ambivalent social behavior, or other forms of psychic disorders by disturbed perception of their bodies (Chapter 3.6). The pectus excavatum deformity with maturation of the female breasts usually leads to their distortion, resulting in a so-called strabism of the breasts (Fig. 1). Usually females affected by such a strabism do not complain about the pectus excavatum deformity itself, which might be partially concealed by the volume of the breast tissue, but rather they definitely dislike this breast strabism, highly focusing the attention of their body perception to such a stain [4].

A method used for the correction of breast strabismus is transposition of the nipple–areola-complex (NAC) to lateral [21] with rotational plasty of the breast volume itself to lateral. A procedure in such a manner may only contingently change the deformity into an improved aesthetic appearance, because herewith the basic and underlying deformity will not be altered, but goes along with additional scars at the breast skin. Furthermore, an augmentation of the breast volume through silicone implants in moderate [23, 24] to severe deformities does not really lead to an improvement of the strabismus, but rather results in a volume augmentation bilateral to the sunken sternum. If now in addition to breast implants a further silicone block is used to simultaneously [6, 22] or sequentially fill up the funnel deformity, this can lead to undesirable results through dislocation of the implants within several confluent tissue pockets (Fig. 2a–c).

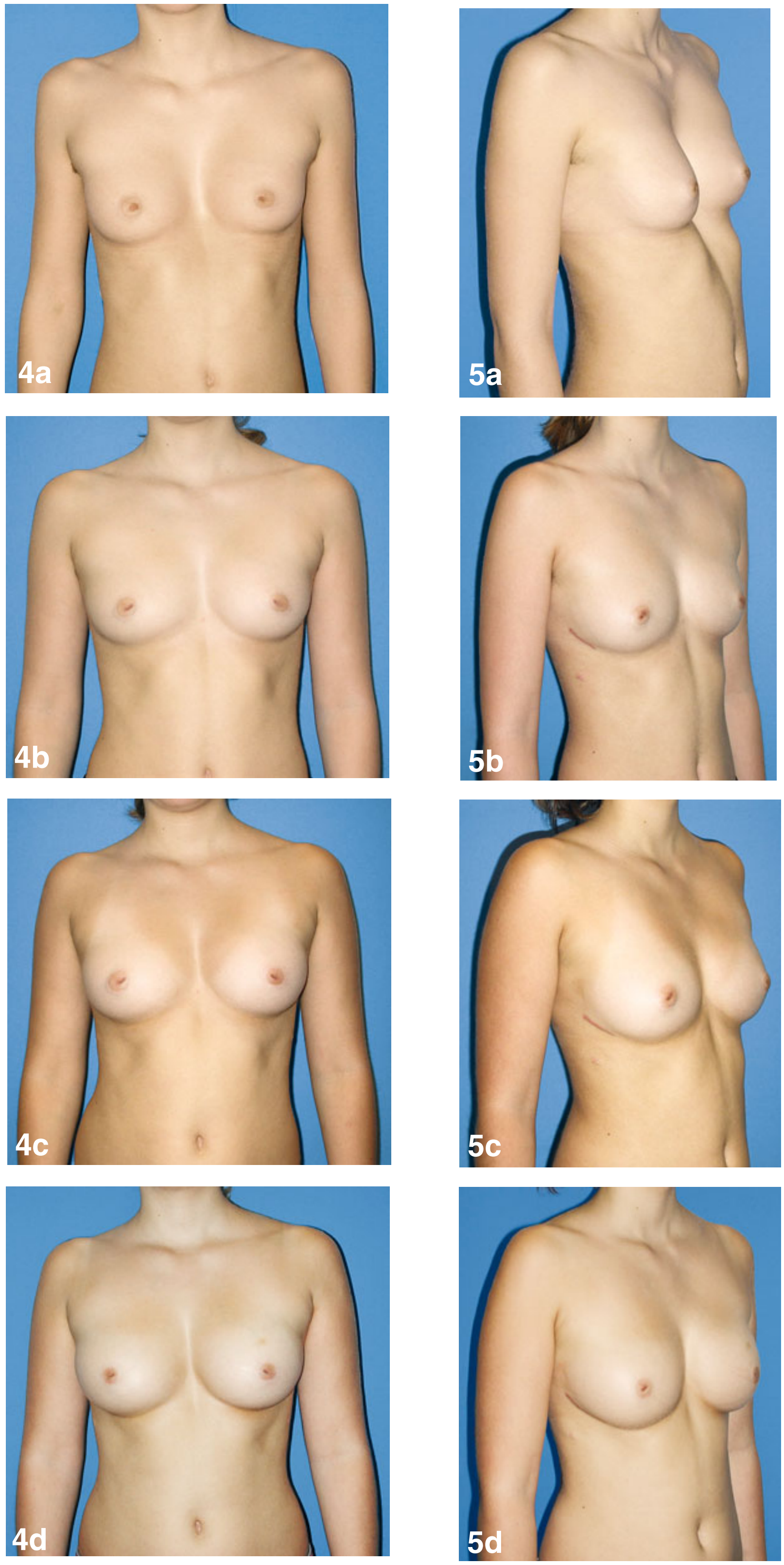
Before surgical methods with a common but unreflected approach are used in order to treat a funnel deformity in females surgically, an exact analysis of the extent of the deformity and the underlying skeletal structures is mandatory. The predominant cause of the strabism does not rely on deformity of the breasts themselves, but rather an inclination of the anterior thoracic wall is responsible for sloping inwards the breast volume. As such the skeletal deformity with the sloping inwards breasts leads to a reduced intermamillary distance, which apparently represents a nipple strabism (Fig. 3a–c CTs). This effect furthermore leads to lacking projection of the medial upper and lower quadrants of the breast (Fig. 3d and e). This deformity being rather a distortion of well-developed breasts therefore corresponds solely to a symptom based on a deformity of the anterior chest wall. However, it cannot be excluded herein, that some of these patients suffer from an additional deformity, whether being breast asymmetry in volume or shape (Fig. 1) as well as hypoplasia or hyperplasia aggravating the dominant chest wall deformity, either in funnel or keel chest deformities. In order to obtain appropriately aesthetically acceptable results, it is necessary to elevate and reshape the anterior thoracic wall to produce a naturally looking position and projection of the breasts (Figs. 4a–g and 5a–g). Minor deformities like an anterior thoracic hypoplasia [24] on the other hand with just likewise neglectable strabism under circumstances can be corrected aesthetically acceptable also with breast implants alone, thus this mainly aesthetic intervention suits very well to camouflage an annoying stain to a considerable extent (Fig. 6a and b).

In some cases, even an asymmetry of the chest wall with malrotation of the sternum exists underlying the breast distortion. Herewith an exclusive augmentation of the breasts with silicone implants can hardly create a satisfying aesthetic result. In rarely existing asymmetry of the breast volume itself conjoined with a pectus excavatum deformity however, it becomes unequally more difficult to obtain symmetry just by augmentation measurements.

By particular modifications of the MIRPE technique with only minor or even without surgical intervention to the female breast tissue itself aesthetically impressive results of the remodeling procedures at the anterior thoracic wall simultaneously with significant improvement of the breasts shape can be obtained.

9.1 Surgical technique of the pectus excavatum deformity in females with chest wall remodeling

The details of surgery are described already in Chapter 6.4. The choice of the surgical method is based on the shape of the funnel deformity as well as on age and general habitus of the patient. In an elastic and symmetrically shaped thoracic cage, with a rather flat depression, the MIRPE method fits ideally for the requirement of chest wall elevation. In patients with sternum malrotation as well as in older patients with skeletal rigidity, the MOVARPE (Minor Open Videoendoscopically Assistet Repair of Pectus Excavatum) modification of the MIRPE method is applied. Herewith a



Preoperatively

6 months postoperatively

14 months postoperatively

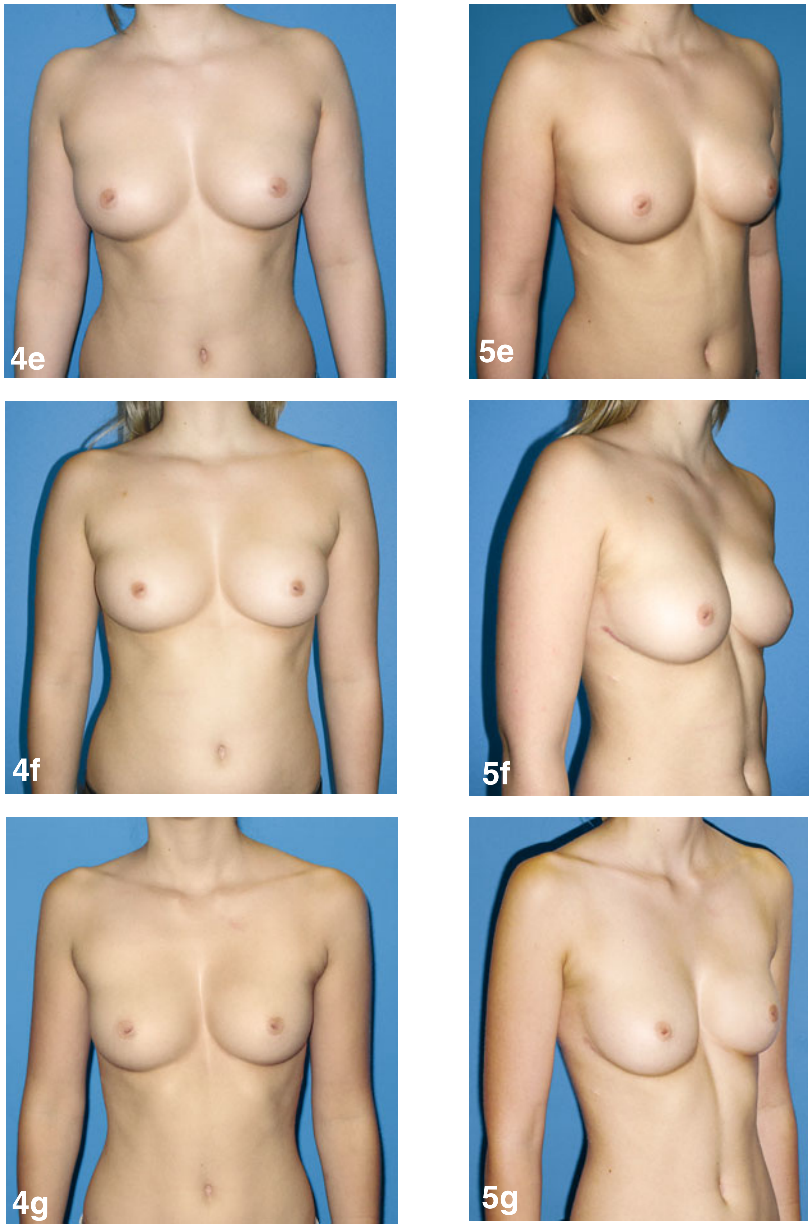
24 months postoperatively

Figs. 4 and 5a–g (*Continued*)

36 months postoperatively

7 months after pectus-bar explantation

36 months after pectus-bar explantation



Figs. 4 and 5a–g. Series of a female corrected with the MIRPE technique at 14 years reaching from the preoperative situation to the situation 3 years after explantation of the pectus bar at 20 years. Notice the slight late relapse 3 years after final surgery but still very pleasing result. Both laterally placed scars are perfectly camouflaged at the submammary crease

horizontal sternotomy is carried out with the access from the submammary crease along a subcutaneous tunnel (Figs. 7a–d and 8a–d). Parasternal chondrotomies or chondrectomies carried out caudal from the level of the sternotomy complete the minor invasive mobilization of the funnel deformity. This hybrid access is necessary on the one hand in order to first enable elevation of the sternum and to put the malrotated sternum into a frontal plane on the other hand. Already intraoperatively, after implantation of the pectus bar, an expansion of the intermammillary distance up to 3 cm is achievable; the strabism is treated therewith by chest wall elevation immediately. With exception in rare cases with a true form of additional volume asymmetry of the breast tissue, no surgery at the breast tissue itself is necessary (Fig. 4). In these rare cases with breast asymmetry however, an adjusting reduction mammoplasty of the greater breast exemplarily using a

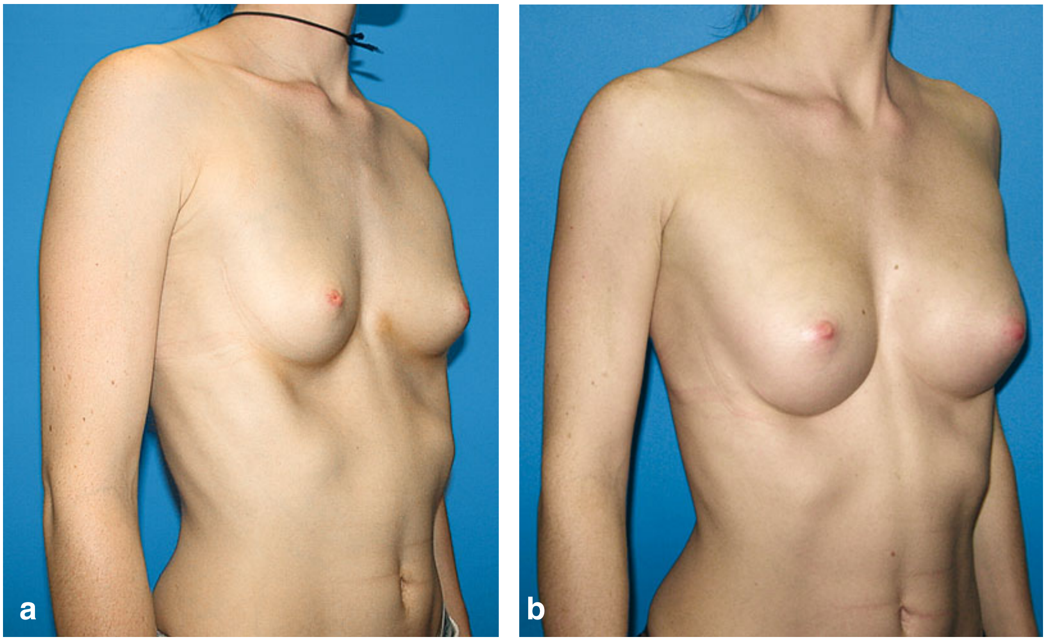


Fig. 6a. 22-Year-old female with a mixed deformity (anterior thoracic hypoplasia) consisting of a minor pectus excavatum but dominant lateral thoracal cavities at the inframammary region. The moderate breast hypoplasia is accentuated by the underlying skeletal deformity. **b** Same patient 1 year after bilateral breast augmentation. This minor invasive (compared to chest wall remodeling) procedure may conceal the chest wall deformity to a considerable thus very satisfying extent in this special case

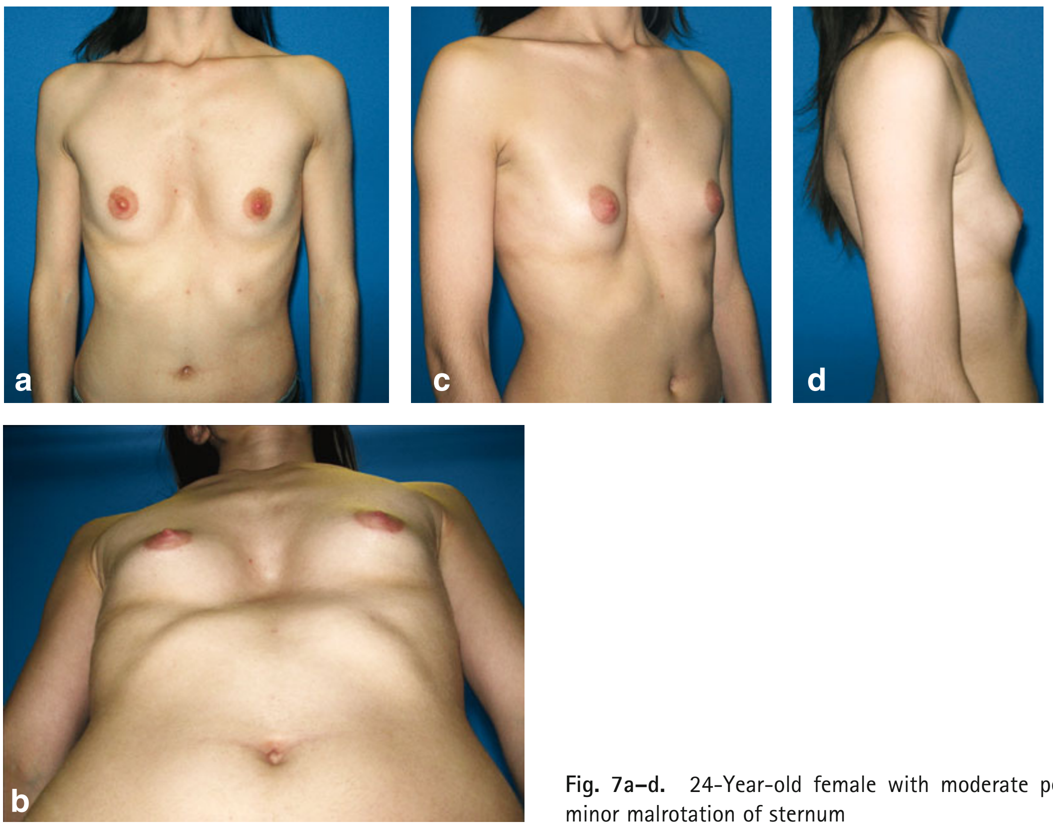


Fig. 7a–d. 24-Year-old female with moderate pectus excavatum and minor malrotation of sternum

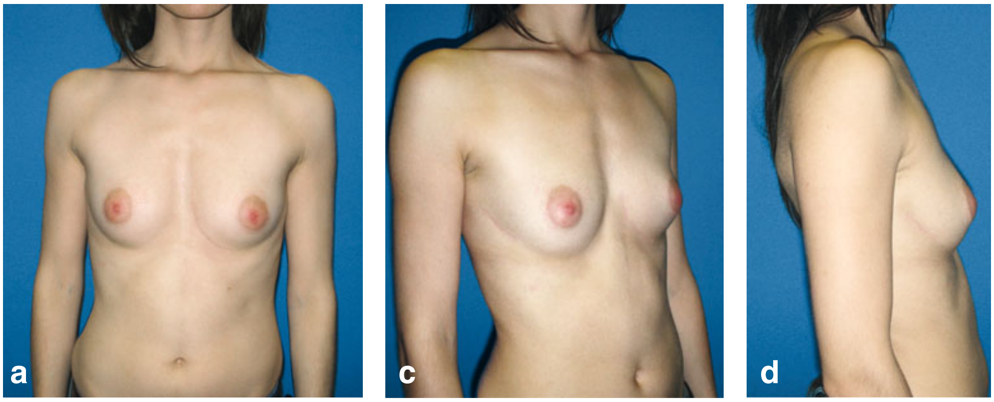


Fig. 8a–d. Same patient as in Fig. 7, 2 years after pectus excavatum repair utilizing the hybrid MOVARPE technique with a left submammary access for sternotomy and parasternal chondroplasties, situation 1 year after explantation of the pectus bar

disc-shaped tissue resection may be carried out exclusively from a submammary incision [12, 19], nevertheless if a pectus excavatum (Figs. 9a, b and 10a–c) or carinatum (Figs. 11a–c and 12a–c) chest deformity represents the basic disorder.

Should an asymmetry exist at the breasts or no satisfying symmetry of the breasts themselves can be achieved by the implantation of one or two pectus bars, adaptive surgery is feasible by reduction plasty at the one larger breast. Nevertheless such a refinement procedure should be undertaken unconditionally first after the period of application time of the pectus bar (Fig. 10c). This is because the breast shape yet can change along the period of pressure dynamics of the pectus bar versus the anterior thoracic wall as the metal strut may dislocate to dorsal with ensuing alteration of the skeletal symmetry.

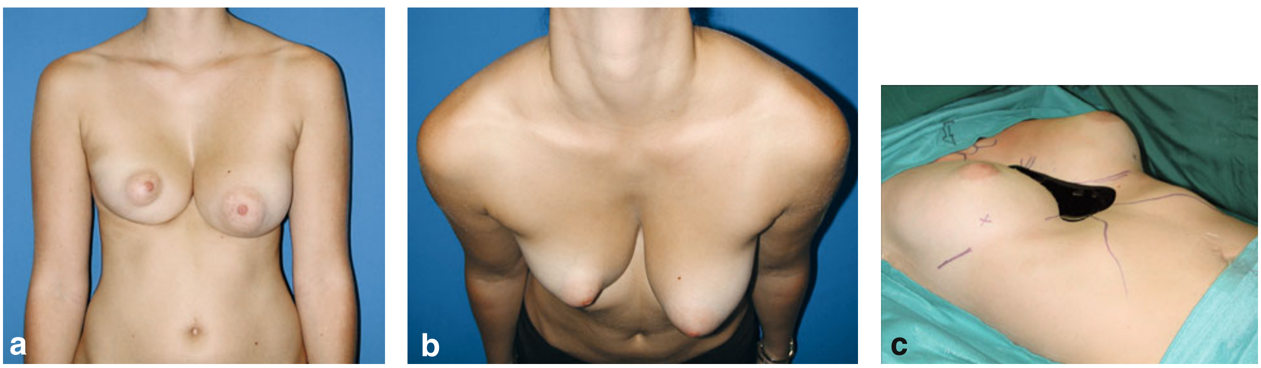


Fig. 9a and b. 20-Year-old female with asymmetric pectus excavatum deformity conjoined with severe asmmetry of the breasts. **c** Intraoperative situs of same patient showing the extent of the chest excavation filled by ink. Lateral marking indicates the skin incision for the pectus bar implantation. Additional incisions at both inframammary creases allow for sternotomy and bilateral chondroplasties to allow elevation of the anterior thoracic wall, then supported by the pectus bar

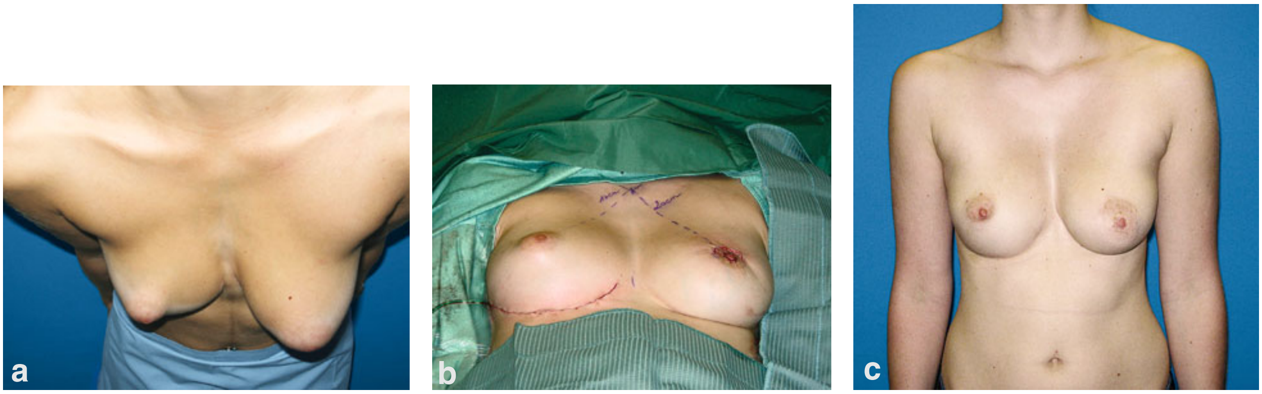


Fig. 10a. Situation 1 year thereafter, prior to scheduled explantation of the pectus bar. Notice the still present asymmetry of breast volume. Volume reduction by disc resection from a submammary access of the left breast was done simultaneously with pectus bar explantation. **b** Refinement surgery with symmetry correction 8 months later by NAC correction at the left side and adaption of the submammary crease at the right side. **c** Final appearance another 1 year later with markedly improved appearance of the pectus deformity and breast symmetry

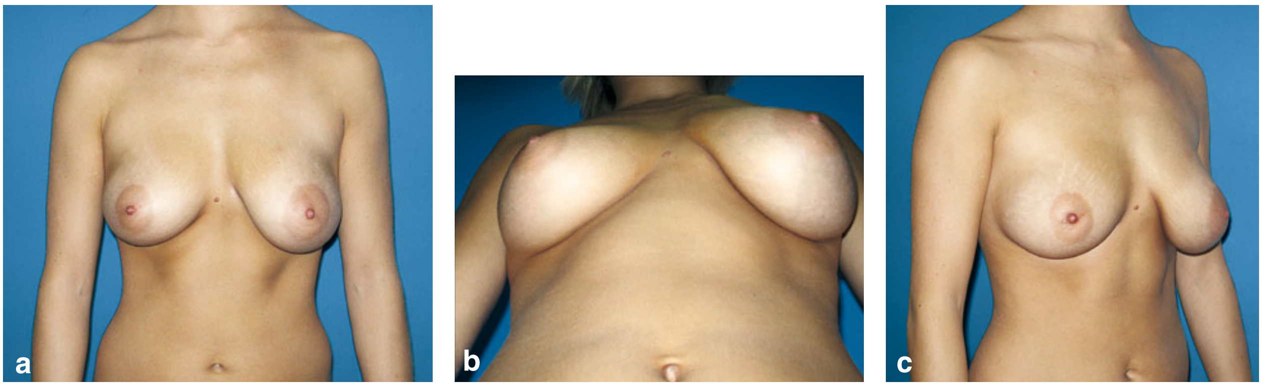


Fig. 11a–c. 21-Year-old female with asymmetry of the breasts, the enlarged breast superimposed on a pectus carinatum deformity at her left side

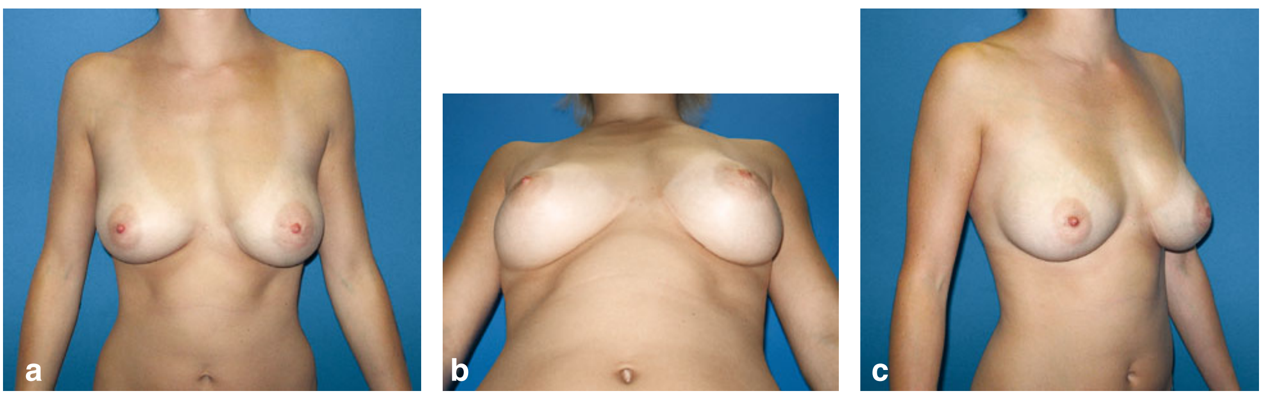
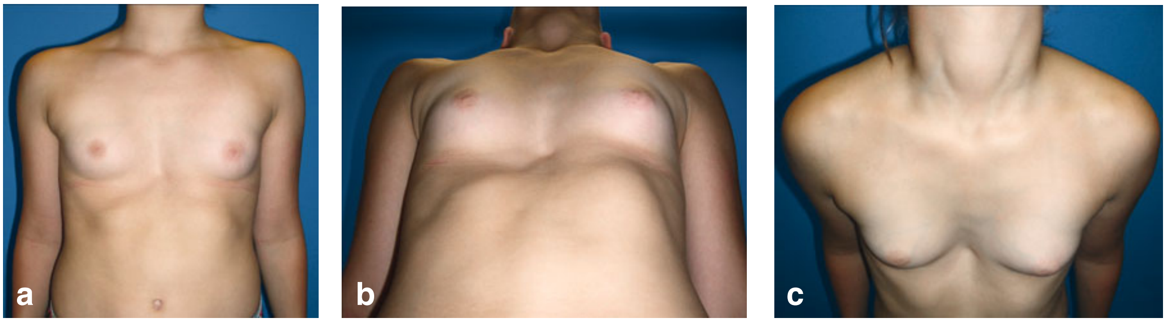


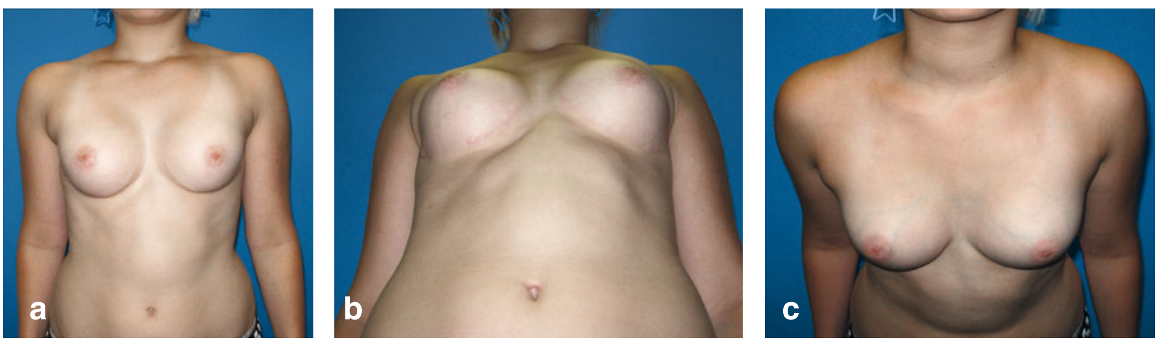
Fig. 12a–c. Same patient 1 year after surgery with only minor remnants of deformed ribs rather cranially. Notice the invisibility of any scar

5 years preoperatively



1 year preoperatively

1 day preoperatively



1 year postoperatively

Fig. 13 (*Continued*)

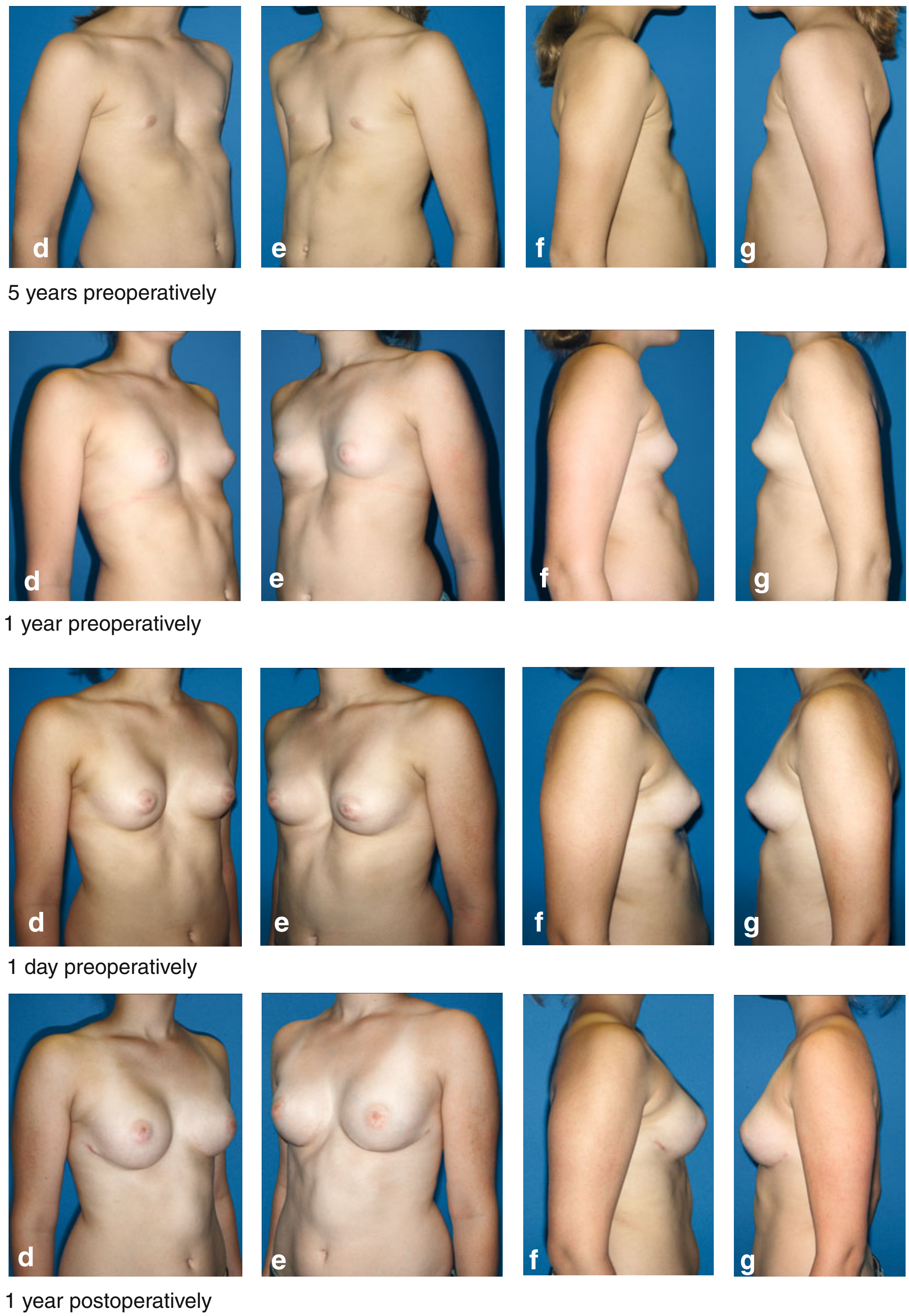


Fig. 13. Series of a young female without functional complaints. The time of pectus excavatum repair was postponed for development of the female breasts in order to place the scars accurately and well hidden. Notice the accentuation of the excavatum deformity over time. The correction was done with the MOVARPE technique using two additional inframammary incisions

Above all in adult patients with strong physique, the pectus bar tends to slip through the intercostal muscles to dorsal if not perfectly placed during implantation. On the other hand through the strong pressure of the sternum transferred via the pectus bar to the levy points at the ribs [7, 13, 26], a deformation of these supporting single ribs with dorsal shifting may cause a slight extent of relapse [11], potentially also asymmetrically. Furthermore, in genuine asymmetrical deformities, such an effect of minor dislocation can result in a secondary form variations of the breasts along the course of several months. Therefore it is advisable to carry out any asymmetry correction in the female breast tissue first along with pectus bar removal after years, when stability of the profile of the anterior thoracic wall can be expected (Figs. 9 and 10). If augmentation due to breast hypoplasia is desired, also such intervention should be postponed and carried out first at the time of pectus bar explantation [2].

9.2 Discussion

Some methods in the correction of the female funnel chest deformity, for example with silicone implants may lead to aesthetically undesirable results. Such undesirable outcome usually is stipulated by weight and gravitational long-term effects, leading to dislocation of the silicone implants, either implated in combination with breast implants or without such. Above all the weight of a major silicone block, placed subcutaneously above the sternum periosteum and adjacent muscles (Chapter 6.5) may lead to the feeling of prepectoral pressure while lying on the back and to unpleasant sensations during sports activities. Moreover such silicone implants are not of lifelong durability so that they are subject of exchange in the course of more or less years due to the development of capsular fibrosis or even capsular calcifications (Fig. 2 in Chapter 12.3). Such implants can correct the deformity of the anterior thoracic wall only symptomatically, therefore do not permit its permanent and durable remodeling.

Through the MIRPE method (Chapter 6.3), worldwide interest and application gained significance in the correction of pectus excavatum deformities, offering major remodeling with only a minimally invasive access via solely lateral scars. These scars especially in females can be placed hidden at the submammary crease inferior to the lateral quadrants of the breasts. Commonly the major collective of patients not until puberty or already adulthood seeks medical consultation for correction of breast deformities. Advantageous in an intervention in puberty or in the young adulthood is the fact that the personality is already ripened and the understanding for this relatively yet invasive intervention is perceived to a better extent, and the compliance with postoperative behavior and treatment is based on the strong desire for a correction, much better than at an early age. Furthermore, this period is advantageous because the female breasts are already well or even fully developed with repect to shape and volume and therewith the submammary creases are well defined (Fig. 13), where scars are supposed to be set. Should an additional sternotomy for the correction of malrotation or deep inclination and rigidity be necessary, an additional skin incision to access this purpose may also be placed very easily into the submammary crease but parasternally (Fig. 9a).

The question whether the intervention, either MIRPE- or the modified MOVARPE technique with an additional thoracotomy would be not too invasive for the purpose of improving a predominantly aesthetic aspect is frequently raised. The experiences during the last years and the frequent use however, above all of the MIRPE showed that with increasing experience the complication rates become less [15]. With corresponding care in the use of these surgical techniques, number and extent of complications may be kept very low

Fig. 14. Minor carinatum deformity bilaterally at the lower part of the sternum with submammary surgical access. The sternotomy also was carried out from the left incision, using an angled oscillating saw (Fig. 3a and b in Chapter 13.2)

Fig. 15a–e. 15-Year-old female with a severe asymmetric pectus carinatum deformity. **f** Same patient with intraoperative markings. The dotted line circumscribes the area of remodeling, the access to it managed by a right submammary incision only. **g** Resected skeletal parts of same patient after osteochondroplasty is completed. **h** Same patient after wound closure, the resulting scar, sufficiently long for adequate remodeling maneuvers does not exceed the submammary crease

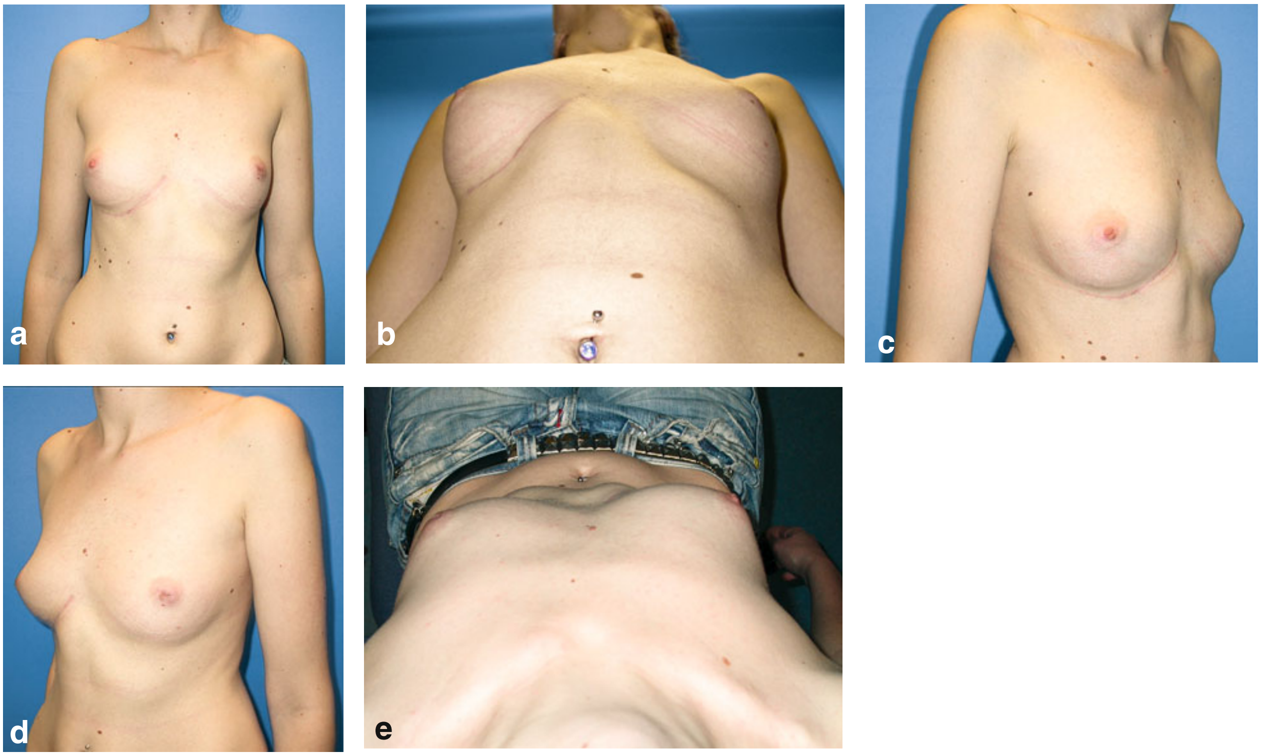


Fig. 16a–e. Same patient 6 months postoperatively

nowadays [8]. It is nevertherless of paramount importance to carry out such interventions, only in central hospitals where corresponding infrastructures are instantly available for the case of major or rarest life-threatening complications appropriately. A correspondingly efficient intensive care unit must also be present.

With regard to these possibilities of embarrassing and in the worst case life-threatening complications, the patient must be informed herewith also correspondingly. The patients seeking predominantly aesthetic corrections with invasive methods must clarify during discussion of the informed consent that the risks have outweighed the potential aesthetic benefit, thus the patient may not be withheld from far less invasive alternative methods such as a silicone implantation or lipofilling interventions (Chapters 6.5 and 6.6.2). Although appealed minimally or minor invasive, the invasiveness of the pectus bar implantation still is considerably high. The term "minimally" stands in contrast to earlier techniques with major invasive tissue dissection at the anterior thoracic wall with ensuing local problems like heavy scarring as an aesthetic or functional impairment.

Fig. 17. Submammary creases elapse while the patient is lying supine at the operation. Manual imitation of gravitational ptosis thus depicting the orthotopic position of the submammary crease exactly where incisions are intended to be placed

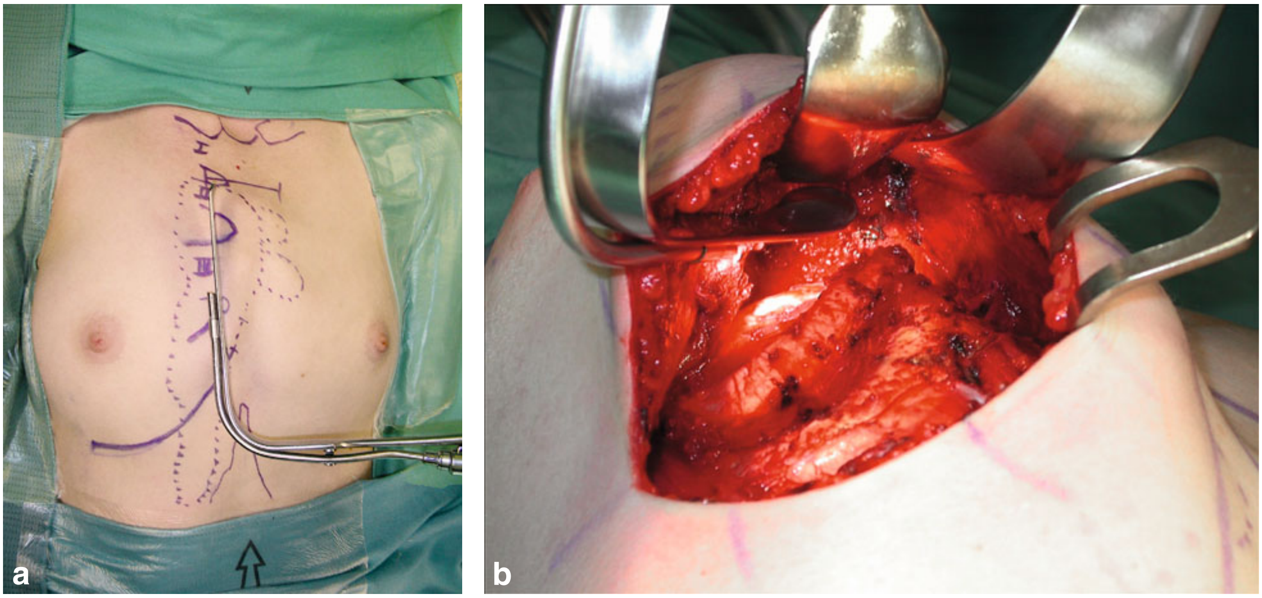


Fig. 18a and b. Specially designed long tissue retractor with attached cold light able to reach the depth of surcically necessary manipulation behind muscles and above distorted ribs

Fig. 19a. Parasternal hump of the second rib at the right side, inaccessible via a submammary incision. **b** In order to avoid scars at the décolleteé, the incision for surgical access to resect the cartilage of the second rib, the incision is placed exactly along the RSTL at the anterior axillary fold, the access the supported by the light equipped retractor as seen in Fig. 18. **c** Situation 6 months postoperatively with a concealed scar and successfully treated rib deformity

In others, than pectus excavatum deformities, the main concern of getting rid of a deformity is the placing of potentially disturbing scars [16]. Whereas in chondrogladiolar deformities the surgical access may be managed via submammary incisions (Figs. 14, 15a–h, 16a–e and 17) and using specialized instruments to reach the site of remodeling (Fig. 18a and b), this is not equally feasible in chondromanubrial or transitional deformities extending more cranially. Herein the incision lines must reach the presternal area to obtain sufficient access for adequate surgical manipulations (Fig. 18a). Minor protrusions at the manubrial area may be reached with special instruments from a lateral access, however the incisions herein have to be exactly placed along the RSTL to avoid unsightly scars (Fig. 19a–c).

9.3 Conclusion

The MIRPE method according to Nuss or the modified MOVARPE version permits a permanent remodeling of the anterior thoracic wall, an aesthetic shaping of the female décolleteé, and the repositioning of the female breast to a physiologic thus naturally appearing position. In most cases repair of the chest wall and repositioning of the female breasts can be accomplished

without surgically touching them. Although such corrective methods are invasive, they raise the quality of life of the concerned patients definitely and therefore to our opinion may be medically indicated in selected cases.
Particular attention must be paid to setting the surgical incisions in younger females with any kind of chest deformity in order to hide the resulting scars at a concealable area, preferentially at the submammary crease. Therefore and if not indicated by functional reasons, corrective surgery should be postponed to puberty, the time when the submammary crease by sufficiently developed breast volume is well enough defined to host scars.

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Special technique in the Poland syndrome

10.1 Surgery of Poland's syndrome

Fazel Fatah

10.1.1 Principles of surgical correction

Planning of surgical correction of the chest wall deformity in Poland's syndrome depends primarily on the severity of the condition and the different types of tissues involved. This may include some or all of the following components: skin and subcutaneous tissues, breast, muscle(s), rib(s), and other skeletal deformities such as scoliosis. The treatment is therefore, tailored to the needs of the patient. The deformity in females has the added dimension of breast asymmetry that may require augmentation or total breast reconstruction. Surgical treatment may involve any or a combination of the following:

- Implant augmentation.
- Custom-made prosthesis.
- Tissue expansion.
- Latissimus dorsi (LD) muscle transposition.
- TRAM flap or other free tissue transfer.
- Lipomodeling, autologous fat graft, and liposuction.
- Rib cage reconstruction with autologous or alloplastic material.
- Nipple areola complex (NAC) reconstruction.
- Reduction, mastopexy or augmentation of contralateral breast.

Although most patients who present for treatment are young adults, increasingly children of the age of 10 years and in the early teens are seeking advice about treatment, but at a much earlier date parents seek advice about the deformity and indications and timing of surgery. Definitive treatment in young males is easier than in females due to the more dramatic changes in the size and shape of the female breast, as they grow older. Men with the deformity and approaching the middle age sometimes seek treatment due to a natural increase in the size of the normal breast and development of gynecomastia with age that leads to a more noticeable gross asymmetry of the chest (Fig. 1a). Mild rib cage deformity and/or partial absence of the anterior segment of one

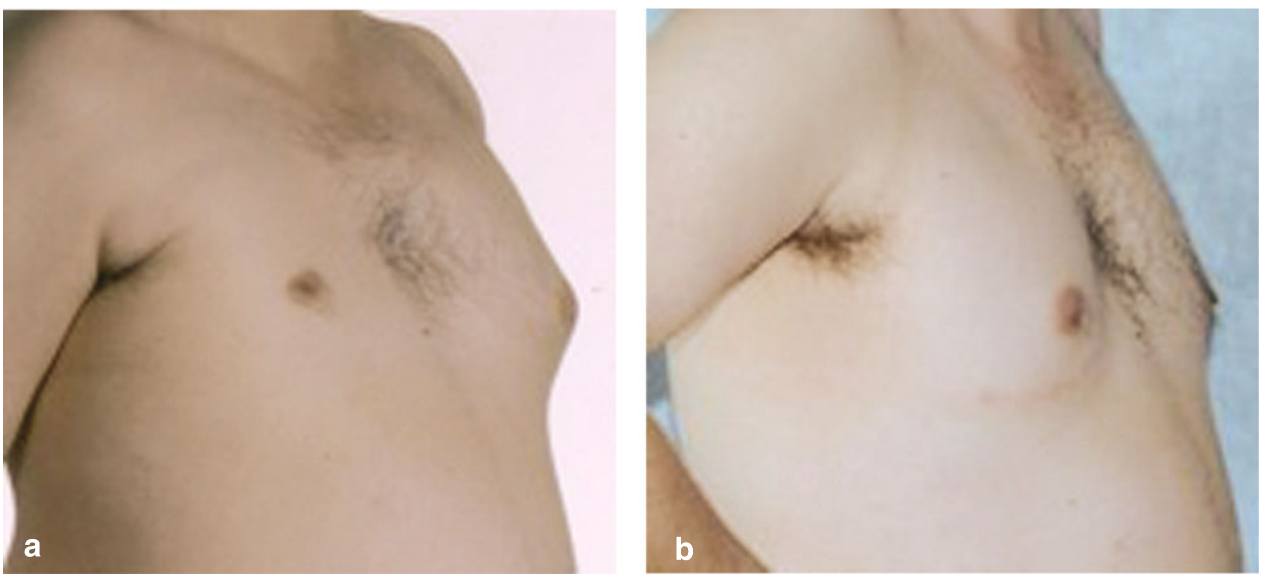


Fig. 1a and b. Correction of Poland's deformity in a 44-year-old male patient using a custom-made silicone prosthesis on the right side and liposuction of gynecomastia on the left side or more ribs, commonly from second to fifth [27], that do not cause an obvious depression in the rib cage contour can be ignored during reconstruction but severe absence of multiple segments of ribs require chest wall reconstruction before the overlying soft tissue deformities can be corrected. Appropriate surgical treatment must be carried out on different age groups in males and females.

Personal experience include treating 32 patients between 1993 and 2007 with an age range of 11–44 years and using a variety of techniques either as single procedures or multistage and combined procedures, examples of which are demonstrated below.

10.1.2 Correction of male Poland's chest wall deformity

Although it has been mentioned that Poland's syndrome affects males more than females [21], in reality, because rarely there is a functional indications for surgery [5], more female patients seek treatment for aesthetic reasons than males do. But increasingly male patients who are self-conscious of the deformity seek surgical correction too.

The method of correction applied depends on the severity of the deformity. Insertion of custom-made silicone prosthesis in a subcutaneous plane can be very effective in camouflaging the deformity [12] (Fig. 1a, b). The consistency of the silicone prosthesis is dictated by the consistency of the tissues it replaces [5]. When the defect is more pronounced and reconstruction of the anterior axillary fold is required, LD muscle transfer can be considered. A custom-made prosthesis may also be required if the contour defect is too severe due to rib cage deformity [12, 15, 25]. However, the patient has to understand that this means sacrificing the posterior axillary fold and adding scars to the back and axilla for the purpose of tissue-addition to the chest. Preoperatively, it is important to clinically establish that the LD muscle on the affected side is equally developed like the contra-lateral normal side [3, 6]. If ipsilateral LD transposition is not available, free microvascular contralateral flap transplantation is an alternative (Chapters 10.2 and 10.3) and it is mandatory to establish the presence of suitable recipient vessels preoperatively as well as intra-operative assessment of the recipient vessels before embarking on the free tissue transfer [4, 20]. The area of muscle deficiency is outlined with the patient standing up and both hands tightly squeezing the waistline for comparison and the skin of the anterior chest wall is marked and extending onto the arm. Similar dimensions are drawn on the back skin overlying the LD muscle as a guide for the dimensions of the muscle harvest required for the reconstruction. Through an axillary dissection the adequacy of the thoraco-dorsal pedicle is ascertained first. The anterior chest dissection is carried out next through an inframammary incision and with the help of a set of lighted retractors. Further access is gained through the axillary incision that is used to identify the pedicle as part of raising the LD flap. Superiorly, in an attempt to reach as high as possible in correcting the deformity in the infraclavicular area, extreme care must be taken to avoid injury to the cephalic and/or subclavian vein particularly if the clavicular head of the LD muscle is absent or hypoplastic. Injury to the subclavian or cephalic vein can lead to disturbing hemorrhage and immediate action must be taken by gaining direct access to the bleeding vein to avoid heavy blood loss. The LD muscle can be mobilized through a relatively small transverse incision in the back, approximately 8–10 cm in length, level with inframammary fold or a vertical incision along the lateral border of the muscle [15] and a 5–6 cm incision in the axilla. A small ellipse of skin can be harvested with the flap and de-epithelialized to bury it under the NAC area to enhance the breast contour (Fig. 2). The muscle is completely islanded on its neurovascular pedicle by detaching the insertion from the intertubercular floor of the humerus and tunnelled through the axilla to the chest and then reinserted to the crest of the greater tuberosity of the humerus using the tendon of the clavicular head as a guide when it is present. Otherwise, in the complete absence of PM muscle, the tendon is sutured to the fibrous condensation of the periosteum of the crest above the deltoid insertion where the latter muscle fibers normally blend with the pectoralis major muscle insertion [15, 23]. This is carried out through the axillary incision and it requires strong retraction of the space between the deltoid and the two heads of biceps muscle. The periphery of the LD muscle is sutured to the chest wall along the lateral border of the sternum and to the lower border of the clavicular head of the PM muscle if it is present. The tendon is inserted to the humerus in a way to create sufficient resting tension in the muscle to continue acting as an effective functional unit and stand out in the anterior axillary fold (Fig. 2c). However, it is difficult to reproduce a fold of similar thickness to the normal side which is formed by rotational twisting of the normal pectoralis major muscle to form a rounded axillary fold before becoming tendinous and inserting to the humerus [23]. This however, can be compensated

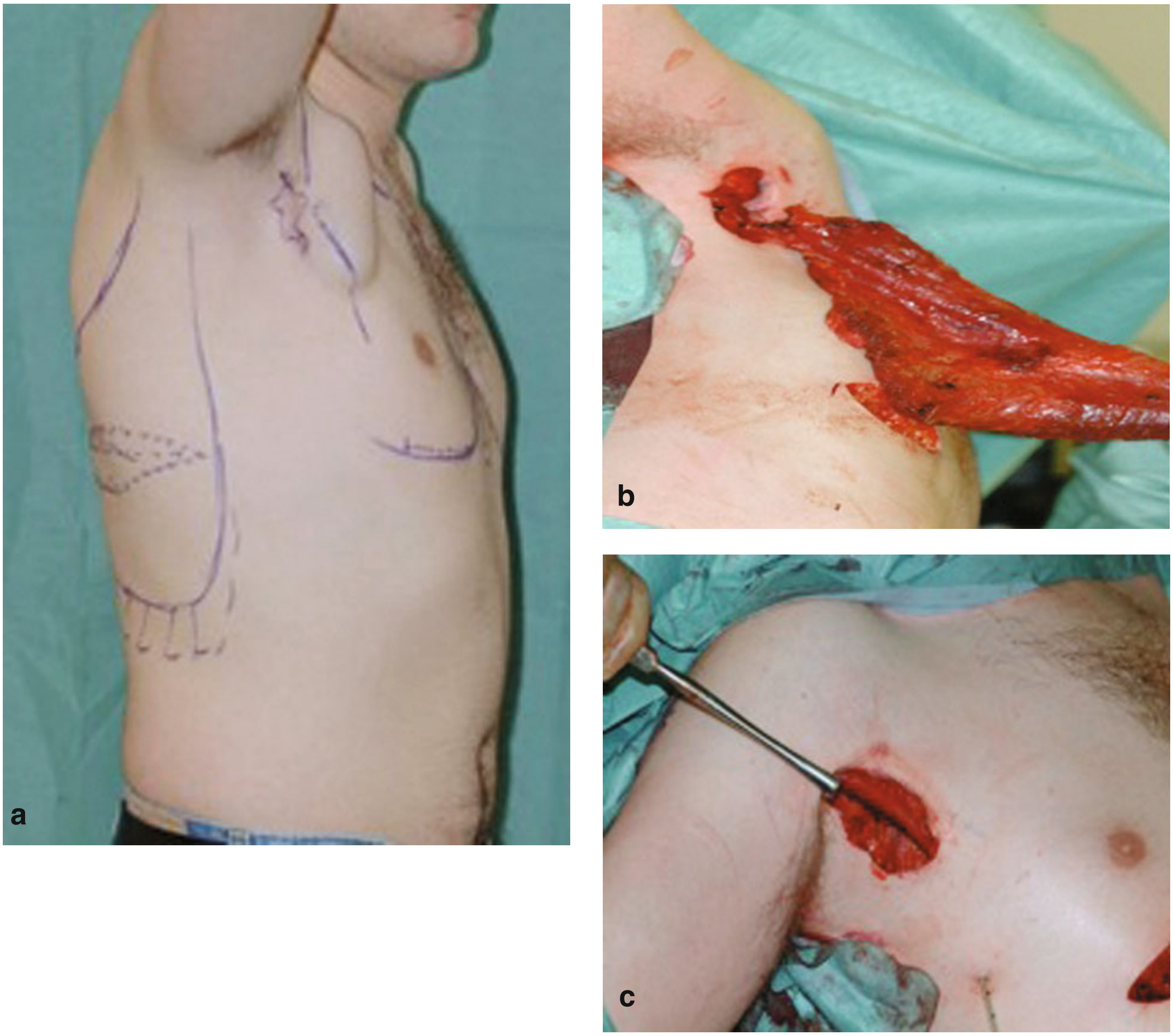


Fig. 2a. Planning of ipsilateral LD muscle transfer. This patient has had prior expansion of the anterior axillary fold skin, notice the –w– scar in the axilla to introduce the expander which is removed and the incision is reused for axillary approach and fixation of the LD tendon to the humerus. Also notice the outline of the dimensions of the muscle required both on the chest and the back including a small skin ellipse to de-epithelialize and use to enhance the breast contour. **b** The LD muscle completely islanded on the neurovascular bundle and ready to be inserted to replicate the PM muscle attachments. **c** LD muscle inserted in a way to reconstruct a functional anterior axillary fold

for with fat injection (Chapter 6.6.2) at a later date if the patient is still concerned. The remaining donor site skin flaps of the back are then quilted down the chest wall with absorbable sutures to prevent seroma formation [10, 24, 26]. In the absence of the ipsilateral LD muscle, the contra-lateral muscle [17] can be used as a free flap (Chapter 10.3). Targeted exercise in the postoperative period can help maintain and even increase the bulk of the muscle. When the skin in the region of the missing anterior axillary fold is unduly tight, tissue expansion can be carried out before the muscle transfer to facilitate the creation of the fold (Fig. 2a).

Lipomodeling is increasingly emerging as a very simple, safe, and effective way of correcting Poland's chest wall deformities, either as a primary method (Fig. 3a–d) or as a secondary procedure to compliment any of the above described techniques that often leave residual contour deficiency and rarely produce full symmetry [7–9, 22]. The procedure is minimally invasive, it does not leave visible scars

Fig. 3a and b. Preoperative views of basic Poland's deformity in a 23-year-old male patient treated with autologous fat injection. **c** 3 months after the first course of fat injection, 110 cc, notice the formation of a new axillary fold. **d** Final result several months after the second course of 80 cc fat injection and some liposuction of right breast with excellent lasting correction of the deformity

behind and it can be carried out as day case surgery. Furthermore, the reconstructed area feels entirely natural with no need for long-term maintenance work. However, the patient must have sufficient excess fat in the proposed donor sites such as abdomen, loins, hips, and thighs. Large amounts of fat may be required and must be injected methodically (Chapter 6.6.2) to correct the contour defect either in one or multiple sessions, using the Coleman system and technique of micro fat grafting to avoid fat necrosis. If there is a significantly tight skin with lack of subcutaneous tissues, staging the fat injection allows the skin to relax after first injecting a relatively smaller amount of fat, 60–100 ml over a wide area, in preparation for further injections at a later dates. Lipomodeling may involve liposuction of the contra-lateral side to help achieve better symmetry (Fig. 1b).

Injectable alloplastic hydrophilic gel materials such as Bio-Alcamid™ (Polymekon, Italy) have been advocated as a simple method for correction of various contour defects including the chest wall and in Poland's deformity [18]. However, the highly viscous material is not easy to inject, particularly when very large volumes are needed to correct such deformities. Furthermore, one has to weigh the relative simplicity of the technique against possible long-term problems that may arise from injecting large amounts of alloplastic material such as infection and granulomatous reactions. Autogenous tissues are always preferable and should be the first choice of treatment whenever applicable.

Asymmetry of position of NAC is more difficult to correct in male patients, however significant asymmetry of size can be corrected with tattooing.

10.1.3 Correction of female Poland's chest wall deformity

The female deformity is more complicated due to the added breast deformity and asymmetry that also involves the NAC. In patients with partial absence of PM muscle and mild to moderate degree of breast asymmetry, unilateral breast augmentation or bilateral differential breast augmentation may be sufficient to achieve an acceptable level of correction. However, implant

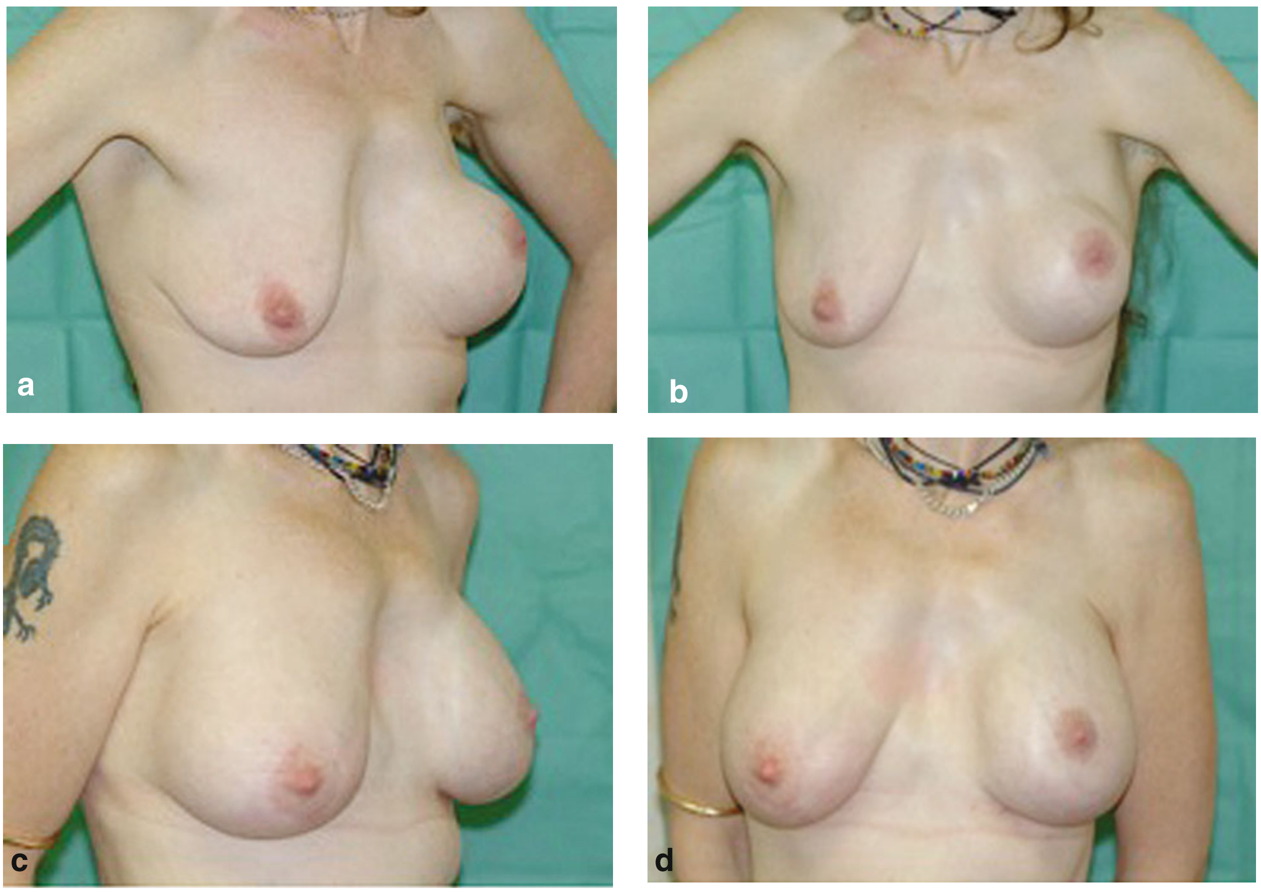


Fig. 4a and b. Unsatisfactory result from attempted correction of Poland's deformity in a female patient using round silicone breast implant. Notice the appearance of an accentuated upper pole deformity and gross asymmetry with her naturally ptotic right breast. **c** and **d** Significant improvement after replacing the old implant with a full height and full projection anatomically shaped (Inamed® BioDimentional) breast implant and mastopexy + augmentation of the normal right breast using a medium height and medium projection of a similar breast implant

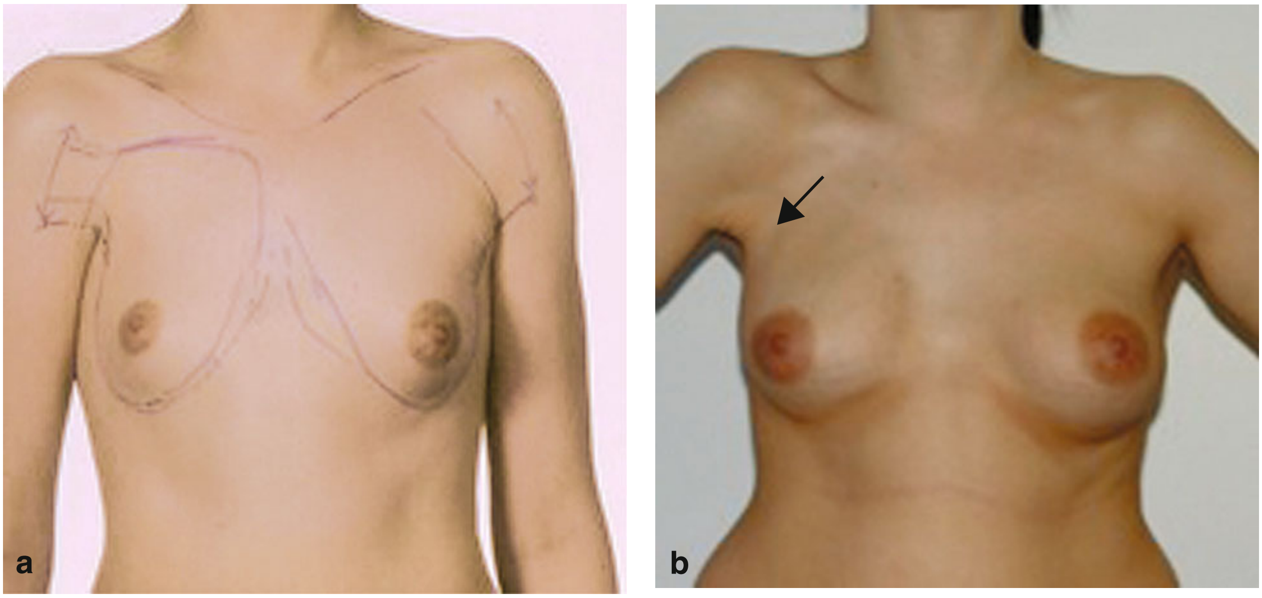


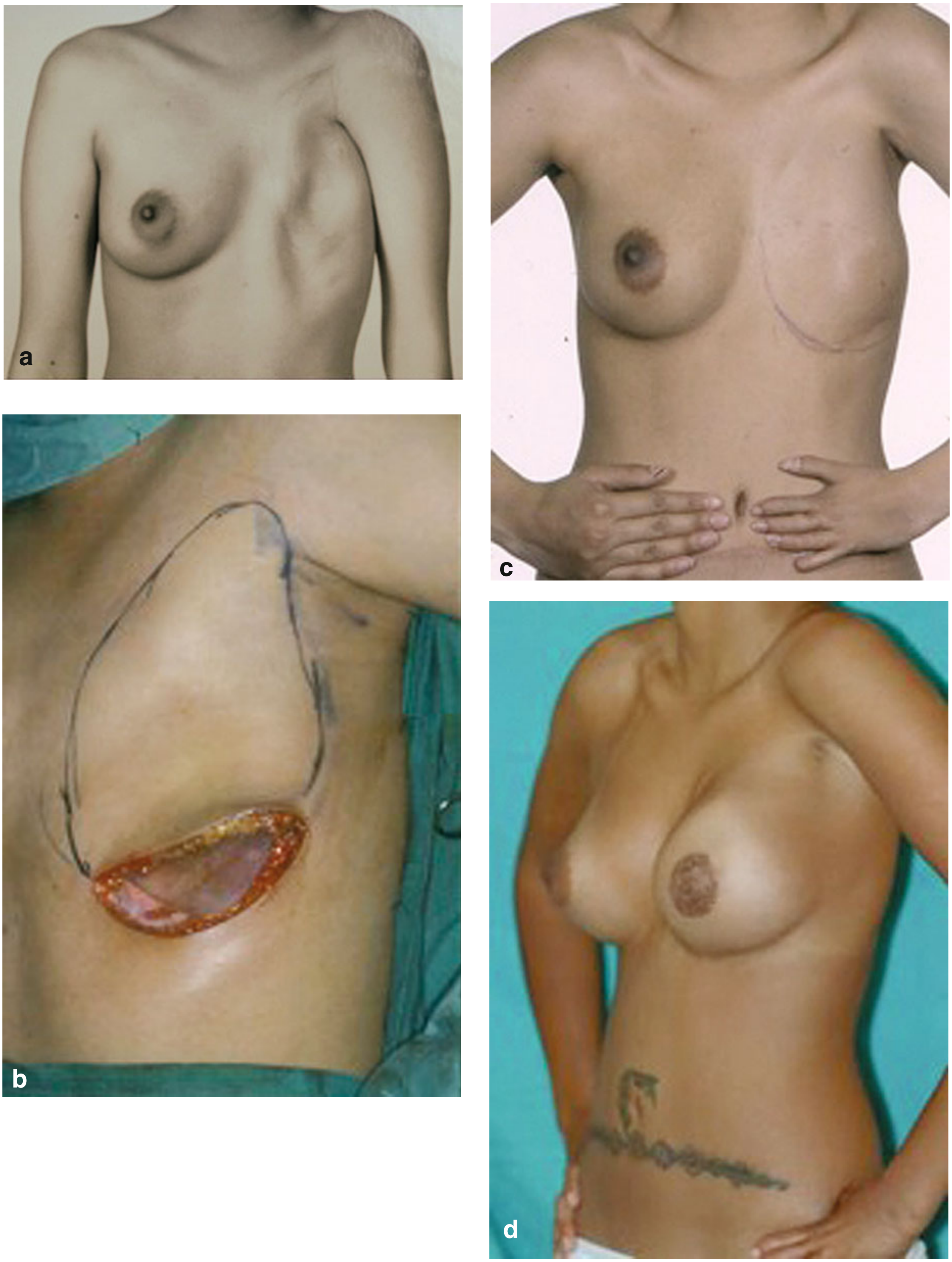
Fig. 5a. 17-year-old female patient with complete absence of right PM muscle and moderate breast asymmetry; planning for a combination of bilateral differential breast augmentation and right LD muscle transfer to reconstruct the anterior axillary fold and reduce the contour deficiency in the infra-clavicular region. **b** Notice a fully functional LD muscle in the right anterior axillary fold indicated by the arrow. Further correction of any residual contour deficiencies can be improved with fat injection

alone does not correct the defects in the infra-clavicular and anterior axillary fold areas. A full height anatomical implant is preferable to help correct some of the upper pole contour deficiency (Fig. 4a–d) [16], which demonstrated a complex reconstruction of the chest wall and breast deformity in a female patient using customized composite silicone prosthesis. Before dissection of the pocket is started it is necessary to establish the integrity of the ribs, as missing ribs also signify missing intercostal muscles (Figs. 1 and 2 in Chapter 10.3), therefore, extreme caution is required during dissection of the pocket to avoid injury to the pleura and the lung. This is highly relevant if one is revising an old implant with capsulectomy and change of prosthesis; the posterior wall of the capsule may effectively be adhered to the parietal pleura. This principle applies to male patients as well if they undergo prosthetic reconstruction.

Breast augmentation can be supplemented by fat injection in the upper pole, the infra-clavicular region and the area of the anterior axillary fold to further correct the contour deficiencies in these areas that cannot be addressed by implant alone [22]. Total correction of significant deformities in females with Lipomodeling, i.e., fat injection alone is possible in selected patients [8, 9].

When breast hypoplasia is combined with tight skin and in cases of breast aplasia, tissue expansion is necessary to create a skin envelope for the new breast [2]. Depending on the circumstances of the case, the expander is either replaced with an implant, or autologous tissue transfer is required either alone or

Fig. 6a. Significant left chest wall deformity in a 19-year old thin patient with a relatively large and fully developed right breast requiring multi-staged procedures. **b** Stage one involved making a cast of the rib cage defect to manufacture a custom-made semi-solid textured silicone prosthesis to create a platform for the breast reconstruction. The prosthesis is introduced in a subcutaneous pocket (see arrow) that was very carefully dissected because of the missing ribs; the dissection was made safer by creating a tumescent field. The prosthesis was fixed with few sutures at the periphery of the rib defects through holes made during the manufacturing of the prosthesis. **c** Stage two involved a bipedicled muscle preserving TRAM flap to harvest a flap with maximum possible dimensions to help introduce some soft tissue in the subcutaneous plane covering the area from below the clavicle to the inframammary fold and from the lateral border of the sternum to the anterior axillary fold. The soft tissue mound is significantly small and further measures needed to create a breast. **d** Stage three, a Style 150 adjustable breast implant was inserted in a pocket dissected between the TRAM flap and the custom-made silicone prosthesis preserving its covering capsule. The implant was inflated to its capacity over a period of several weeks and in a stage four, nipple reconstruction was carried out and later followed by tattooing of the NAC. The whole process took approximately a year to complete and as a final touch the patient went and had a decorative tattoo designed by a professional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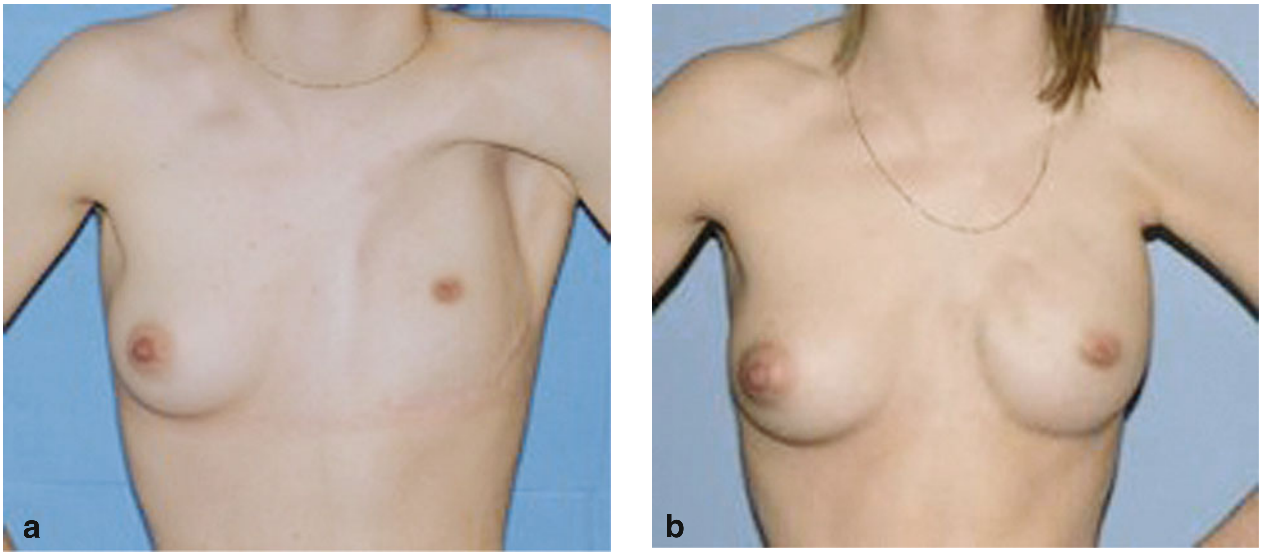

Fig. 7a. Rib cage deformity corrected in the cardiothoracic department using methylmetacrylate cast introduced through an incision near the inframammary fold region and the patient referred for breast reconstruction. **b** Breast reconstruction with a combination of LD muscle transfer and Style 150 full height anatomical implant resting on a platform of methylmetacrylate prosthesis. Notice the functional LD muscle in the anterior axillary fold

combined with an implant. LD muscle transfer is ideal for this purpose with the implant inserted behind it. The muscle contributes to the correction of the upper pole and the anterior axillary fold contour deficiencies as well as providing added soft tissue cover for the implant (Fig. 5a, b). Alternatively, Lower abdominal excess tissues can be transferred either as a pedicled TRAM, free TRAM, DIEP or SIEA or other free perforator flaps [11, 13, 20]. In the presence of severe absence of multiple rib segments with obvious chest wall defect and severe breast hypoplasia or aplasia (Fig. 6a), appropriate reconstruction of the skeletal defect is necessary before the breast and other soft tissue defects can be rectified. The reconstruction becomes more challenging when the patient is thin and has a well-developed contralateral breast (Fig. 6a). Preference should be for the least invasive technique with the minimum visible scars and complications. Custom-made silicone prosthesis is preferable and serves this purpose well. Alternatively, methylmetacrylate cast made at the time of the exploration can be used (Fig 7a). In a second stage soft tissue transfer is carried out. If LD muscle is used, an adjustable expander/implant can be inserted under the muscle at the same time to be expanded at a later date (Fig. 8b). However, if TRAM or other autologous free tissue transplantation is carried out, the flap is de-epithelialized and inserted under the skin with attention paid to correcting the upper pole and anterior axillary defects which takes a significant volume of the flap to achieve. Insertion of the expander/implant under the transferred tissues is deferred to a later date to prevent problems with flap survival due to pressure (Fig. 6b–d). A narrow strip of skin from the flap can be left and inset in the skin incision made in the future inframammary fold to monitor circulation in the flap that could be removed at a later date. This technique avoids insertion of a skin paddle from the flap as in delayed breast reconstruction in order to create natural ptosis. If the patient has a high NAC on the affected side, a strip of skin from the flap can be inserted above the NAC to lower its level. Further procedures to reconstruct NAC and make final adjustments of shape and size may be necessary.

10.1.4 Correction of Poland's chest wall deformity in children

Children between the ages of 8 months and 16 years have been treated for chest wall deformity including the use of LD muscle transfer, ipsilateral as well as contralateral micro-neurovascular transfer, rib grafts, and synthetic mesh. So far, no evidence of long-term adverse effect on the musculo-skeletal development is reported [1, 14, 17, 19]. However, one must consider the fact that the procedure is primarily carried out for

Fig. 8a. 11-year-old girl had insertion of Style 150 implant after becoming very self-conscious of her relatively minor breast asymmetry and absent pectoral head of PM muscle when she was 12 years old. At this stage the augmented breast is larger and no saline insertion was needed for another 2 years. **b** Same patient at the age of 18 requiring change of implant. **c** Now the patient is aged 21 and notice the change in her weight, she requested change of her implant in the right breast for a bigger one

aesthetic rather than functional reasons [5] and perhaps unless there is a compelling reason for operating earlier in life [1], one should allow the child to grow to an age when they can demonstrate signs of self-consciousness and willingness to undergo surgery. Young girls with minor form of the deformity commonly become more aware of it once they begin to develop a normal breast on the contra-lateral side which can happen as early as the age of 10–11 years. Many of them are happy to postpone treatment until they have matured further and some until they have fully grown. If they wish to have early treatment, it is reasonable to use a suitable adjustable breast implant with potential to increase the volume as the contra-lateral breast grows larger (Fig. 7a–c) until such time when a more definitive treatment can be carried out with the patients' full informed consent. The earlier surgery is carried out the more likely that revisions will be required, as the patient grows older, particularly in female patients.

In conclusion, Poland's chest wall deformity includes a wide range of visible deformity and asymmetry, from simple to complex, depending on the severity and the extent of the different tissues involved. Rarely treatment of the chest wall deformity is required for functional reasons. A variety of treatment options are available, ranging from simple to very complex and multistage procedures. There is no age or sex limitation on surgery as such, but judgment must be exercised in each patient on an individual basis, taking the patients' specific circumstances into consideration including the presence of other associated deformities.

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10.2 Special microvascular flap for the Poland syndrome, the TMG-flap

Thomas Schoeller

10.2.1 Introduction

The Poland syndrome deformity is characterized by the missing lower part of the pectoralis major muscle and therefore the missing anterior axillary fold (Fig. 1). Additionally hypoplasia of the breast gland can be associated in various forms. Severe other deformities of the upper limb can be also seen and are described in a different chapter (Chapter 2.3.4). The idea of correcting a missing pectoralis major muscle by a free functional transverse myocutaneous gracilis (TMG)-flap is a relatively new approach to this particular problem. In former times the most common surgical correction was the transposition of the latissimus dorsi flap, if this muscle was not effected by the Poland syndrome [1–4]. In some centers lipofilling is used as a primary method of correction [5]. In our hands the lipofilling procedure is more used as an ancillary procedure for minor volume corrections (see Chapter 6.6.2). The TMG-flap gives the opportunity to create a dynamic and functional anterior axillary fold by inserting the gracilis muscle in the direction of the missing pectoralis major muscle followed by a nerve coaptation. Additionally the attached skin can be used to build up the missing soft tissue volume in the infraclavicular region and if necessary also to add further volume to the missing breast.

10.2.2 Surgical technique

The operation is performed in two teams, one is preparing the recipient pocket (Fig. 2) for the gracilis-flap in the pectoral region of the affected side, the second team simultaneously harvests the TMG-flap at the contralateral thigh (Figs. 3–5). The skin is undermined from the thoracic skeleton and the recipient vessels, preferably the thoracodorsal vessels, if available, as well as a recipient motor branch from muscles in the vicinity (Fig. 6). Due to the characteristics of the Poland syndrome a high variability of unexpected anatomy occurs with sometimes, apart from the absent pectoralis muscle and breast gland, hypoplastic or even missing further anatomical structures [6].

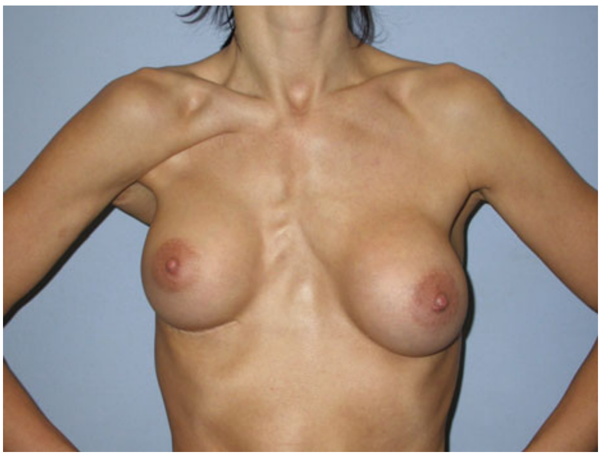

Fig. 1. Missing anterior axillary fold due to hypoplastic pectoralis major muscle (PMM). Only the clavicular portion of the PMM is visible during forced contraction. This patient already received subglandular breast implants in a former operation

Fig. 2. Marking of the recipient pocket in a subcutaneous plane to host the transplanted microneurovascular flap. The orientation of the pocket simulating the direction of the lacking portion of the PMM

Fig. 3. Marking of the gracilis muscle along the thigh, with a transverse skin island along the groin and the anterior gluteal fold, in order to keep the resulting scar in a hidden region

Fig. 5. Harvested free transverse myocutaneous graciilis (TMG)-flap with the vascular pedicle (right) and nerve (45° angle to vessels)

Fig. 4. The flap already dissected, notice absence of any scar at the thigh. The gracilis muscle was dissected via the skin incision, that circumcised the skin island

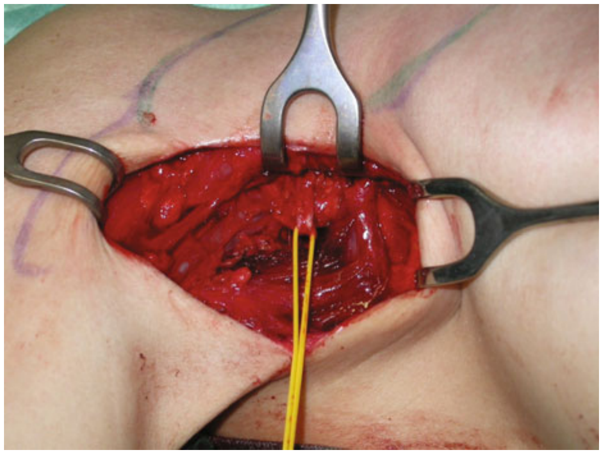

Fig. 6. Thoracodorsal nerve marked with a rubber loop, to which the anterior branch of the obturator nerve supplying the gracilis muscle will be coapted end to side

10.2.3 Harvesting of the TMG-flap

The patient lays in supine-position, the thigh is slightly abducted and the calf mobile, trapped with sterile stockings. The skin island of the flap is featured by its landmarks which are: the upper border is the natural crease of the groin in extension to the infragluteal fold (Fig. 3). The anterior border is the palpable tendon of the adductor longus muscle, the posterior endpoint is the posterior midline and the lower boarder of the skin island is defined by the laxity of the skin which will be tested preoperatively in a standing position. The skin is pinched and approximated to the upper line where tension free closure is still possible. Usually a width of the skin island of 8–12 cm can be harvested. The skin island is elevated including the fascia of the thigh, posterior and anterior of the attachment of the gracilis muscle. The skin island after harvesting remains attached to the gracilis muscle only. In the next step the plane underneath the facia and above the gracilis muscle is mobilized along the whole length of the inner thigh. Attention is paid to the secondary minor pedicle of the gracilis muscle. In many cases direct clipping of this pedicle can be achieved, but if this pedicle arises very distal only a blunt dissection with a slowly overextension and subsequent rupture of the pedicle may cease bleeding therefrom. As soon as the conjunction between tendon and muscle is felt, the tendon is dissected with long scissors, but transected as far distally as possible. The origin of the muscle will be released from the pubic bone and the nerve motor branch and the vascular pedicle are prepared up to the length requested. The motor branch (anterior branch of obturator nerve) should be followed as proximally as possible (up to 10 cm is rather feasible). The motor branch enters the muscle about 2 cm above the hilus of the vascular pedicle (Fig. 5). The nourishing vascular side branches into the adductor longus and abductor magnus muscle have to be clipped off and the pedicle is further developed until it originates from the profunda femoris artery and vein. The closure of the donor side resembles a typical medial thigh lift. Skin closure by resorbable sutures is recommended since stitch removable in that region is very uncomfortable for the patient. Postoperative compression stockings to the thigh is necessary for the first 10 days.

10.2.4 Preparation of the donor side

Preoperative markings are defining the submammary fold and a symmetrical approach to the new anterior axillary fold (Fig. 2). Just posterior to the new anterior

Fig. 7. The free flap is transferred to the site of reconstruction, depicting the volume that is available with this flap, necessary to fill up the missing tissue and occasionally depressed thoracic wall

Fig. 8. The situation immediately after wound closure. An oxygene probe serves for flap perfusion surveillance. The plastic pads secure transcutaneous fixation sutures to keep the muscle and skin flap at the position desired

axillary fold a lazy-S skin incision is made and the skin at the anterior thoracic skeleton is undermined together with the breast tissue, if present. Special care has to be taken by that step because sometimes ribs are missing [6] and perforation of the thinned thoracic wall may cause pneumothorax (Chapter 10.3). As soon as the whole area is undermined the volume filling flap is inserted (Fig. 7). Prior to flap insertion the skin island needs to be deepithelialized entirely. The subcutaneous part is used to fill up the missing volume either at the infraclavicular region or may serve to add further volume to an underdeveloped breast in women. Fixation of the flap to the deep tissue layers especially toward the midline is facilitated by special hooks equipped with cold light. Then the anterior axillary fold is created by suturing the tendon of the gracilis muscle to the humerus bone or the upper arm fascia with particular attention not to damage any crossing nerves. The origin of the gracilis muscle then is sutured to the rib periosteum and intercostal muscles to the same position as the contralateral pectoralis major muscle presents itself (Fig. 8). Care has to be taken to maintain the correct resting tension, which preferably should be marked with a suture into the muscle before release of the tendon during flap harvest. As soon as the whole flap is sutured and secured, the vascular pedicle will be hooked up by microsurgical anastomoses. End-to-end-anastomoses to the thoracodorsal vascular bundle are preferred. An epineural window to the thoracodorsal nerve is cut, followed by an end-to-side neurorrhaphy secured with a drop of fibrin glue. At the recipient side no compression should be applied after wound closure.

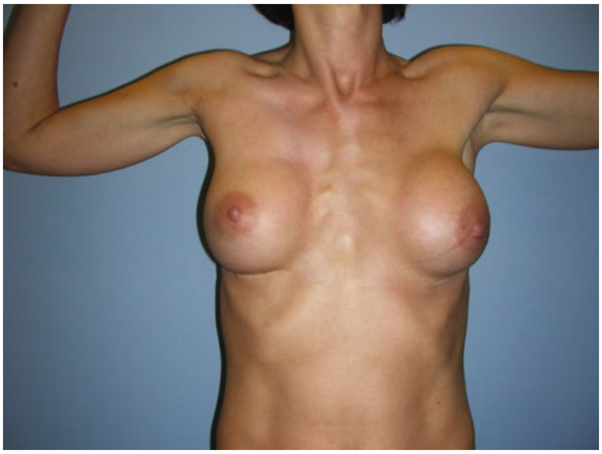

Fig. 9. The situation one year after flap transfer. Notice the sufficient volume bulk at the upper implant margin and the recreated anteror axillar fold at the right side. The reconstructed side resembles even more aesthetic than the contralateral side with a lateralized breast implant due to slight capsular contracture

10.2.5 Discussion

The TMG-flap reconstruction for a Poland syndrome associated thoracic wall deformity is a new and dynamic approach to this difficult problem. It has to fulfill two tasks: first the restoration of the anterior axillary fold and second a volume replacement (Fig. 9). The idea behind the nerve coaptation and the functional transfer is not to provide strength as an adjunct to an underdeveloped pectoralis major muscle, but should create a natural looking and contracting muscle bulk at the anterior axillary fold. Usually half a year thereafter EMG-records show first positive signs of muscle flap reinnervation. Therefore it is of paramount importance to maintain the normal resting tension during surgery, otherwise the muscle fibers cannot contract properly. Depending on the discrepancy of the breast volume and the available tissue at the medial thigh and buttock, the skin paddle can be sufficient to restore a missing breast volume also. Optionally an additional silicone breast implant may be necessary just to adapt the breast volume to symmetry. A second TMG-flap for the missing volume can be inserted if a pure autologous reconstruction is intended and silicone implants are refused. If only a small volume discrepancy has to be corrected, lipofilling would be the method of choice for refinement ([5] or Chapter 2.3.4).

10.2.5.1 Advantages

The major advantage of that procedure consists in the relatively inconspicuous donor site at the thigh which can be placed in a concealed natural body fold. The loss of the gracilis muscle function is totally compensated by the remaining adductor muscles and therefore not really noticeable for the patient [7, 8]. The three-dimensional shape and volume of the TMG-flap allows a lot of freedom for the surgeon to insert the flap depending on the individual requirements of the deformity. The TMG-flap provides with the potential for a dynamic and functional reconstruction and all the benefits of an autologous tissue reconstruction [9–11].

10.2.5.2 Disadvantages

Since the TMG-flap transfer is a microvascular procedure all the risks associated with microanastomoses have to be taken into consideration. Therefore patients with a general contraindication for microvascular procedures should be excluded from that procedure. It bears the risk of a total failure in case of an anastomosis problem. If not trained routinely for other indications, it may become a very long and tedious procedure. Regarding the donor site sometimes sensory branches from the posterior thigh nerve cross very superficial into the skin island and have to be dissected during incision along the skin island in the posterior part of the TMG-flap. This might result in a numb area on the posterior aspect of the thigh. However, and in contrast to the harvest of an infragluteal flap, it will not result in a disturbing neuroma, which has to be mentioned to the patient in the informed consent preoperatively.

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10.3 Special microvascular flap for the Poland syndrome, the latissimus dorsi-flap

Anton H. Schwabegger

10.3.1 Introduction

As an element for the reconstruction of the anterior axillary fold in the Poland Syndrome with partially or completely missing M. pectoralis major, the ipsilateral latissimus dorsi as a muscle (LDM)-flap or eventually as myocutaneous-flap offers itself as a transposed-flap ([1–3] and Chapter 10.1). In some cases however, at the ipsilateral side an associated deformity with hypoplastic or even missing components of the thoracic wall, namely of ribs, intercostal muscles and also of the Latissimus dorsi muscle [2, 4] may be found (Figs. 1 and 2). Apart from such muscle and skeletal anomalies, also the probability exists that moreover yet vascular anomalies in the sense of hypoplasia or aplasia of the thoracodorsal vessels, which are necessarily important for a transposition, are present. As already described by Beer in 1996, prior to a planned transposition of the LDM it is absolutely necessary above all to perform a detailed functional test and suitable imaging (e.g., angio-MRI) of the latissimus- and skeletal thorax region in regard to the vascular situation of the concerned side [4]. If significant anomalies should exist, which indicate lack of sufficient circulation, other alternatives like autologous tissue transplantation or lipofilling ([5] and Chapter 6.6.2) must be taken into consideration for smaller corrections, or microvascular transplanted flaps (Chapter 10.2) for major defect reconstruction. But also in planned microvascular transplantation of autologous tissue, an exact clarification of vascular anatomy must take place in order to test the situation of the recipient vessels whether namely as nourishing vessels they show sufficient diameters and course length for anastomosis of a muscle or myocutaneous-flap or if they are present at all. Furthermore a three-dimensional-CT imaging (Figs. 1 and 2) is extremely helpful in order to be able

Fig. 1. Three-dimensional volume rendering CT scans reveal hypoplastic and fused ribs I–IV. The intercostal spaces are narrowed, as a sign of lesser function due to hypoplasia. The internal thoracic artery of the affected side seems of much lesser caliber than contralaterally. Conjoined with the underdeveloped skeletal morphology this means an indirect sign, that also the thoracodorsal vessels might be hypoplastic at that side

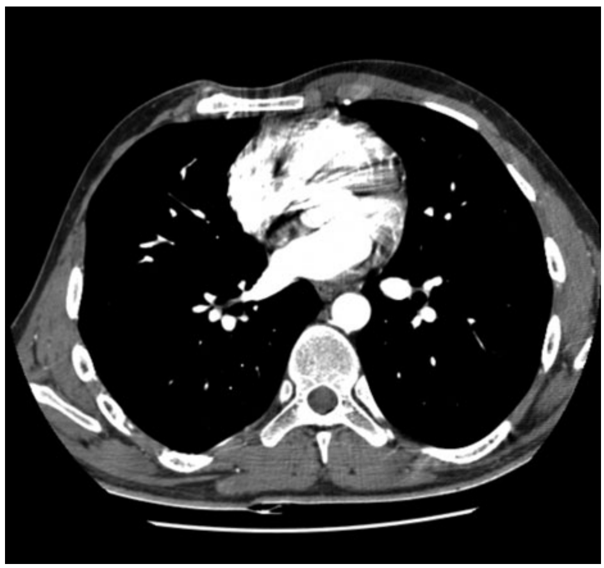

Fig. 2. Conventional CT scan reveals absent pectoralis major muscle, absent ribs and extremely thinned soft tissue separating skin and pleural cavity at the right side

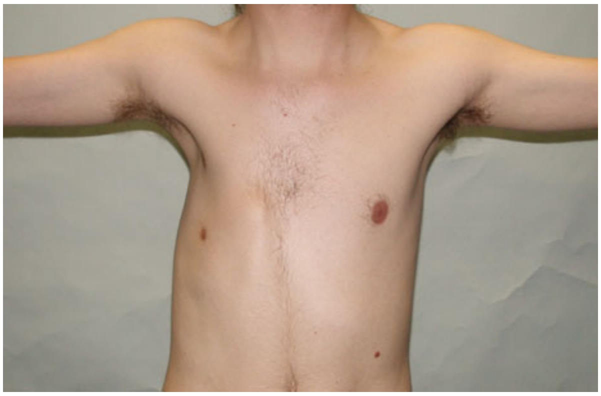

Fig. 3. A 25-year-old male with Poland syndrome, normal anatomy of the left upper extremity but partially absent pectoralis major muscle at the right side

Fig. 4. Additionally a parasternal asymmetric keel chest deformity at the affected right side is clearly visible from an oblique aspect, extending from ribs IV to VII

to detect any possible hypoplasia at the lateral thoracic wall itself. Such anomalies can cause a keel (Figs. 3–5) or funnel chest deformity wall at their variants, however there can also exist merged, hypoplastic, or several missing ribs. In this particular situation, i.e., if no further tissue sheet is present between subcutaneous tissue and pleura, the danger of damage to the pleura and lungs thoroughly exists during the surgical preparation of a tissue pocket as a recipient bed for the flap to be transplanted. The danger of such pleura damage is relatively high also along a Lipofilling procedure, because herewith the fatty tissue is injected with more or less sharpened cannula and in multiple layers into the subcutis (Chapter 6.6.2). This danger of unintended perforation is imminent when muscles and ribs are absent, and also because during anesthesiologic ma-

Fig. 5. Associated displacement of a hypoplastic nipple-areola complex (NAC) into the axilla, even accessory and minor NAC is present therefrom caudally

chine respiration the intrathoracal pressure is raised thus keeps the pleural layers very closely to the adjacent thin skin.

The microvascular transplantation of a Latissimus dorsi muscle was mentioned by Hester already in 1982 and by Beer in 1996, the microneurovascular, i.e., with nerve coaptation was described by Kelly [6] as a case report in 1999. While in females the transverse myocutaneous gracilis (TMG)-flap as described in Chapter 10.2 very well suits not only for a recreation of the anterior axillary fold but furthermore also for augmentation of a hypoplastic or even missing breast, the LDM as a pure muscle-flap ideally can serve as replacement for a missing PM (pectoralis major) muscle because of its similar fan-shaped anatomy. Dependent upon the desires, requirements and possible other concerns with regard to function loss and scar development, the contralateral LDM may be transplanted as a microneurovascular flap, when an ipsilateral LDM-flap is not existing, hypoplastic, or with uncertain blood supply.

10.3.2 Surgical technique

The technique concerning recipient site and flap harvesting differs from that in the TMG-flap procedure. Simultaneous tissue pocket preparation and flap harvesting is not possible, because the patient has to be positioned left or right sided alternately. As already mentioned in the previous chapter, unexpected anatomy, if not properly investigated prior to surgery may cause severe problems concerning revascularization of the transplanted flap and may lead to complications like unintended perforation into the pleural space [6].

10.3.3 Harvesting of the LDM-flap and preparation of the donor side

This technique of flap harvesting is only shortly described here and not in detail, as it is a common and standard procedure in reconstructive surgery worldwide. First the recipient side is prepared in lateral position. Even if in the preoperative imaging the anatomical situation appears clear with sufficient vessel calibers, it is very advisable, first to prepare the recipient site, in order to anticipate any possible anatomical variants and in order to have sufficient time at disposal for possible intricate or time consuming preparations. The preparation of the tissue pocket is sufficiently described already in Chapter 10.2. First if the anatomical situation is assured and the recipient tissue pocket is prepared, the patient can be turned onto contralateral side for the purpose of harvesting of the LDM-flap. It is of paramount importance to maintain ischemia time of the muscle-flap to be transplanted very short, especially if the muscle-flap will be transplanted as a functioning microneurovascular flap, i.e., with coaptation of the thoracodorsal nerve to nerves at the recipient side. An ischemia time up to 2 h is well tolerable, after 4 h already half of the muscle fibers will develop necrosis and after 6 h of ischemia, complete necrosis of all muscle fibers and therewith fatty degeneration will take place without possibility of any reinnervation and therewith total loss of contractile muscle function. The longer ischemia time lasts, the more edema will develop within the transplanted muscle. In the reconstruction with the LDM solely as a muscle-flap within a subcutaneous tissue pocket, in whose preparation the overlying skin must be stretched, an edematous swelling muscle would choke its own blood supply, in the sense of a compartment syndrome. In addition to the probabilities of prolonged problems during performance of the vascular anastomoses at the site of reconstruction, it is therefore imperative, that ischemia time between flap raising/harvesting and revascularization must be held as short as possible. Under certain circumstances, e.g., with slim patients, raising and harvesting of the LDM may easily succeed with the patient laying at his back, the shoulder elevated and the thorax slightly turned utilizing suspension pads [7].

Fig. 6. Postoperative aspect after correction of the keel chest deformity and microneurovascular LDM transplantation from the contralateral side and nerve coaptation to the intercostal nerve III

Fig. 7. Good result of parasternal contour after correction of the keel chest deformity through a parasternal incision according to the Ravitch technique and postoperative keel chest device compression. This operation was performed one year prior to the LDM transplantation

After carrying out flap harvesting a partial or on demand eventually complete relocation of the patient takes place, so that insertion of the flap with microsurgical anastomoses and nerve coaptation is comfortably possible. According to the anatomical situation, the thoracodorsal nerve will be coapted to Intercostal nerves, to pectoral muscle branches or other available motor nerves in an end-to-end or end or end-to-side fashion. As already mentioned, particular care has to be taken into account to maintain the correct resting tension, which preferably should be marked with a suture at the muscle surface before release of the tendon and/or muscle origin during flap harvest [8].

Fig. 8. The NAC from the axilla was transplanted and extended to a symmetrical size to match the contralateral side; the accessory NAC was excised

Such a maintained resting tension then supports an aesthetically pleasant result of the axillary fold as well as an occasional function of a replaced pectoralis major muscle (Figs. 6–8).

10.3.4 Discussion

A microneurovascular autologous transplantation succeeds with functional acceptable results all the more sooner the earlier it is carried out in the youth of the patients [6]. Not only the reinnervation in the pediatric population is significantly better, also the distensibility of the skin in children generally is superior to adults.

10.3.4.1 Advantages

The particular advantage of the LDM (the healthy, contralateral side) exists in its constant and very well-known anatomy as well as in a vast experience with this tissue-flap in reconstructive surgery in general. Moreover it substantially shows larger calibers and a longer vascular pedicle in contrast to most other flaps available or suitable for the actual purpose. In analogy and additionally also the thoracodorsal nerve, which is a requisite for the purpose of a reinnervation of a functioning muscle, provides with constant anatomy and usually sufficient length. For the case that additional tissue volume is required, it can be lifted also as a myocutaneous-flap with a correspondingly large skin island [9], with or without deepithelialization. The more fan-shaped structure and muscle fibers anatomy correspond that of the missing pecoralis major muscle (PMM), so that out of this aspect the LDM appears ideally suited as replacement or recreation of the PMM.

10.3.4.2 Disadvantages

In analogy to all other microvascular procedures also here the risk of circulation problems with partial or entire flap loss exists, however substantially much lesser compared to other flaps due to its constant anatomy and extensive overall experience. Harvesting of the contralateral muscle-flap on the one hand can lead to weakening of retroversion of the arm, but on the other can reduce the prominence of the posterior axillary fold and therefore can create shape symmetry, assimilating to the usually hypoplastic anatomy of the side concerned by the Poland syndrome. The donor site scar at the back is well visible while in males preferably it is supposed to be put laterally onto the thoracic wall. It is more eye-catching in females and not that easily concealable, as the donor site region of a TMG-flap at the groin and thigh region.

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Surgery of other congenital anomalies of the anterior thoracic wall

11

11.1 Isolated rib deformities

Anton H. Schwabegger

Isolated rib deformities in the sense of skeletal prominences may characterize minimally pronounced forms of keel chest deformities, which are caused by overgrowth of a single rib or several conjoined ribs. Such a deformity can occur unilaterally parasternally (Figs. 1 and 4), but may also be present bilaterally and symmetrically (Figs. 2 and 5). Above all the symmetrical rib deformities at the area of the lower thorax aperture can mean a sufferable aesthetic deformity for many patients. Such deformities almost always and without exception present without functional impairment. Occasionally patients complain about pressure symptoms however based on such bony or cartilaginous prominences, above all in the prone or lateral position. Occasionally there are also job-related troubles through carrying special security belts, which must be put around the thorax. Such deformities, if do not lead to job-related functional impairment or troubles must be regarded as pure aesthetic deficiencies, representing solely as a subjective impairment. This type of deformities most frequently is located in the area between 7th and 10th rib and hereby around the transition of the cartilaginous to the bony part of the rib. Particularly here they are sometimes well visible especially in slim patients and during forced inspiration or elevation of both arms (Fig. 2a). The patients apart from an aesthetic flaw complain the symptom complex of pressure pain in the prone and at times also in the lateral decubitus position, which may then indicate a surgical correction. The surgical therapy of such isolated rib deformities or rib humps is rather straightforward because no particular invasiveness for the remodeling is necessary. The skin incisions are put along the RSTL so that after healing these scars are hardly visible any more after months (Fig. 3a–c). However, especially in the depth, within which through separation of the musculature to reach the skeletal structures special attention must be paid on the directions of muscle fibres of the different muscle groups located here. In the paramedian area the rectus abdominis muscle passes along a longitudinal cranio-caudal axis. Therefrom laterally the horizontally irradiating segments of the serratus anterior muscle is found. Further caudally and laterally is yet found the obliquus externus abdominis muscle which passes in an oblique direction from cranial to caudal. Ideally these muscles are then split bluntly along the direction of their muscle fibres (Chapter 7.2), in order to circumvent any damage of functioning muscle units [10]. After depiction of the underlying cartilaginous and osseous rib parts a subperichondrial and/or subperiosteal resection of abundant length of these structures is undertaken in analogy to the technique described by Ravitch [8]. After shortening the ribs, the length of the opened perichondrium and periosteum tubes is shortened with long-term absorbing or non-absorbable reefing sutures. Prior to that special attention must be put on the resection margins of the rib cartilage and rib bone. Ideally these have to be rounded off before the reefing sutures are applied (Chapter 7.1). A careful reposition-

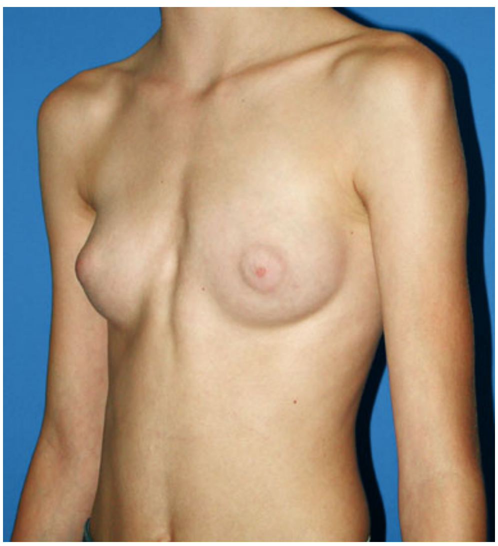

Fig. 1. 15-Year-old girl with isolated rib humps parasternally at the 5th and 6th rib at the left side and a minor neglectable hump at the right side

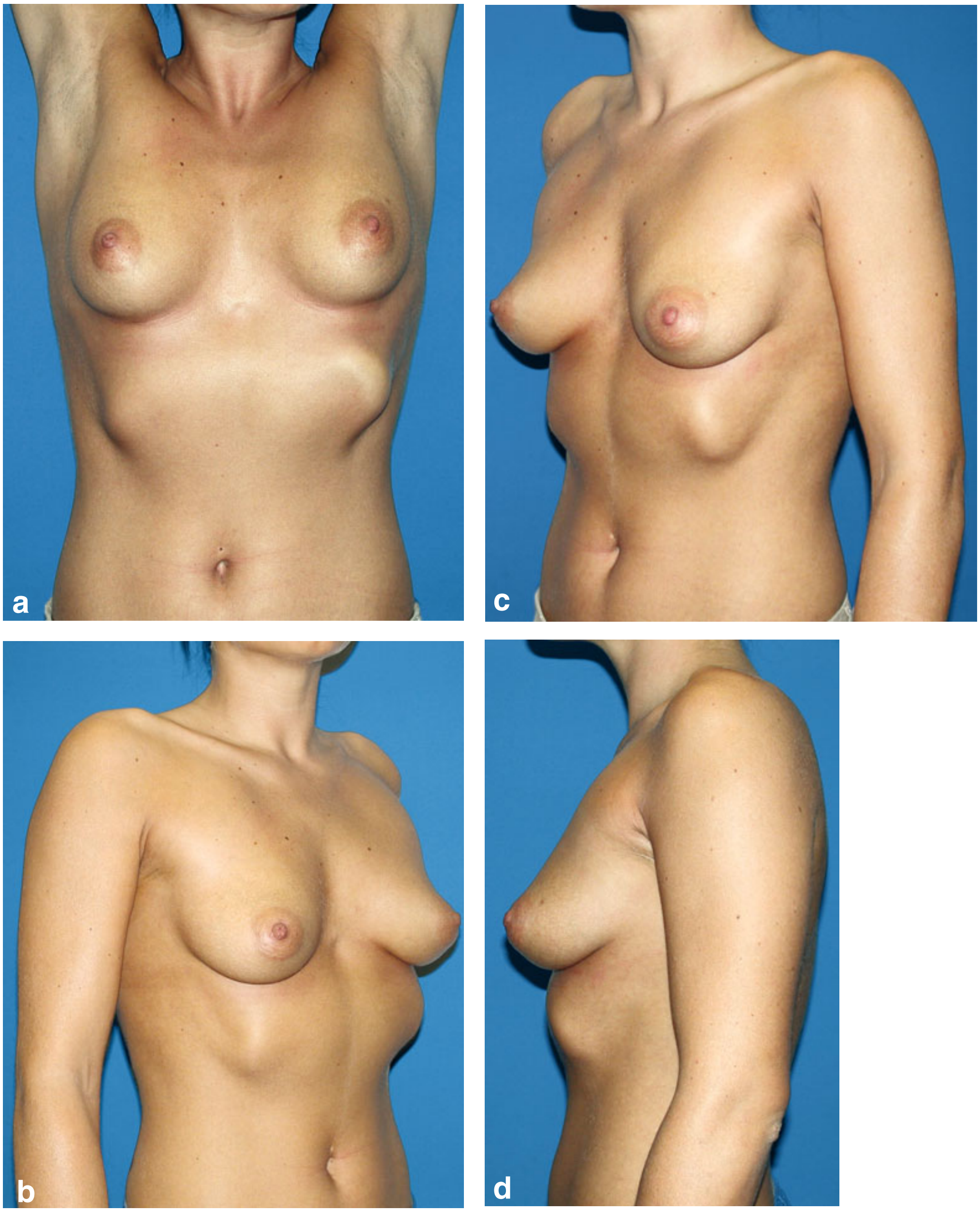

Fig. 2a–d. Preoperative situs in a 24-year-old female with bilateral rib hump deformities prominent at the lower rib arches

ing of the deflected muscles above the site of rib resection and reefing sutures has to be accomplished in order to avoid visibility or palpability of these interventions to the skeletal structures.

Particular attention on the intercostal nerves is also mandatory, because in the lateral thoracic area, where such rib humps or prominent rib arches usually occur, they run along the subcostal groove (sulcus costalis) and present with rather stronger calibres than parasternally. Their damage during rib dissection or during application of reefing sutures in that region may cause cumbersome and long-lasting intercostal neuralgia on the one hand, or on the other hand permanent anesthesia.

After careful resection and likewise careful surgical treatment of the osseous and cartilaginous resection margins actually no tight reefing sutures are required to achieve an improved contour of the thoracic wall.

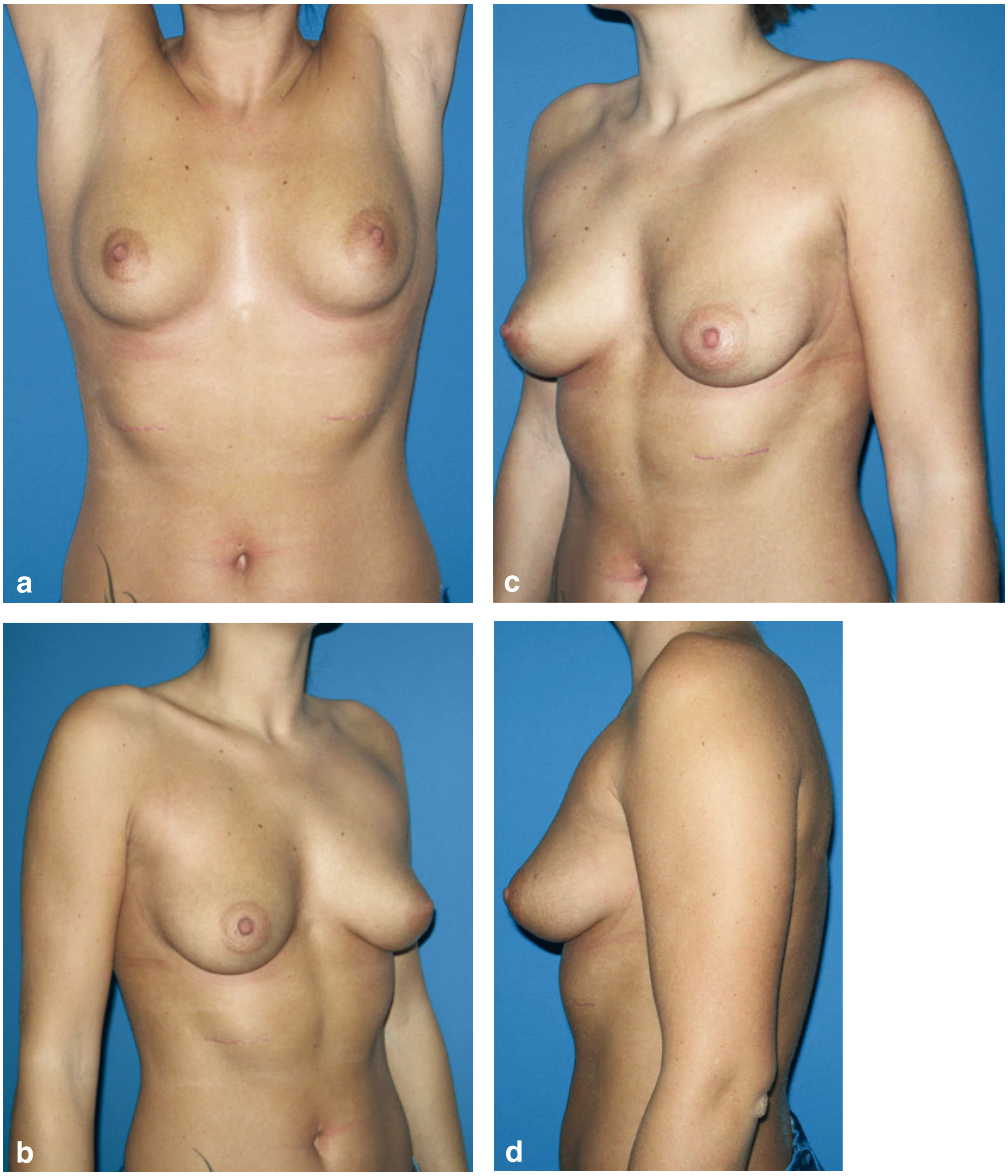

Fig. 3a–d. Same patient 6 months after surgery with resection of the chondo-osseous junction zone of the 7th to 9th rib bilaterally. Skin incisions were set exactly along the RSTL

This is in contrast to the chondrectomies and chondrotomies that are performed parasternally in different deformities, as typically executed in the genuine keel or funnel chest deformities. Because in a concerned area the contour of isolated rib deformities or rib humps is bent and misshaped in a three-dimensional manner, showing a curved course in a frontal, sagittal, and longitudinal plane, a reefing solely along one axis would be feasible only with very high mechanical strain. Such efforts will not succeed and will inevitably lead to recidivation through the elastic properties and memory of artificially bent rib parts, after resorption of or pulled out sutures. It is therefore more advisable to resect the concerned rib parts on a longer range and to be radical to achieve a harmonious height balance, which is then more optimally adapted to the surrounding level of the thoracic wall [9].

Such isolated rib deformities may occasionally be associated with other deformities, particularly with funnel or keel chest deformities or the Poland's syndrome. Particularly in extensive pectus excavatum deformities such prominent, compensatory vaulting of the lower thoracic arches may be present. If then such a funnel deformity is elevated using a pectus bar, such preexisting deformities gain prominence, based on the elevation of the whole anterior thoracic wall. Yet through the maneuver of the remodeling of the pectus excavatum deformity, another deformity, namely a prominence of the rib edge arch will even be accentuated. Such a newly created deformity may vanish after some months through the effect of muscle traction but may also persist thus annoying the patients permanently. Because of optional self-regulation through altered biomechanical strain caused by the pectus bar and muscle traction over a period of months, such deformities should not be corrected at the initial surgery. However, if persistent, they may be corrected simultaneously at the time of planned pectus bar explantation.

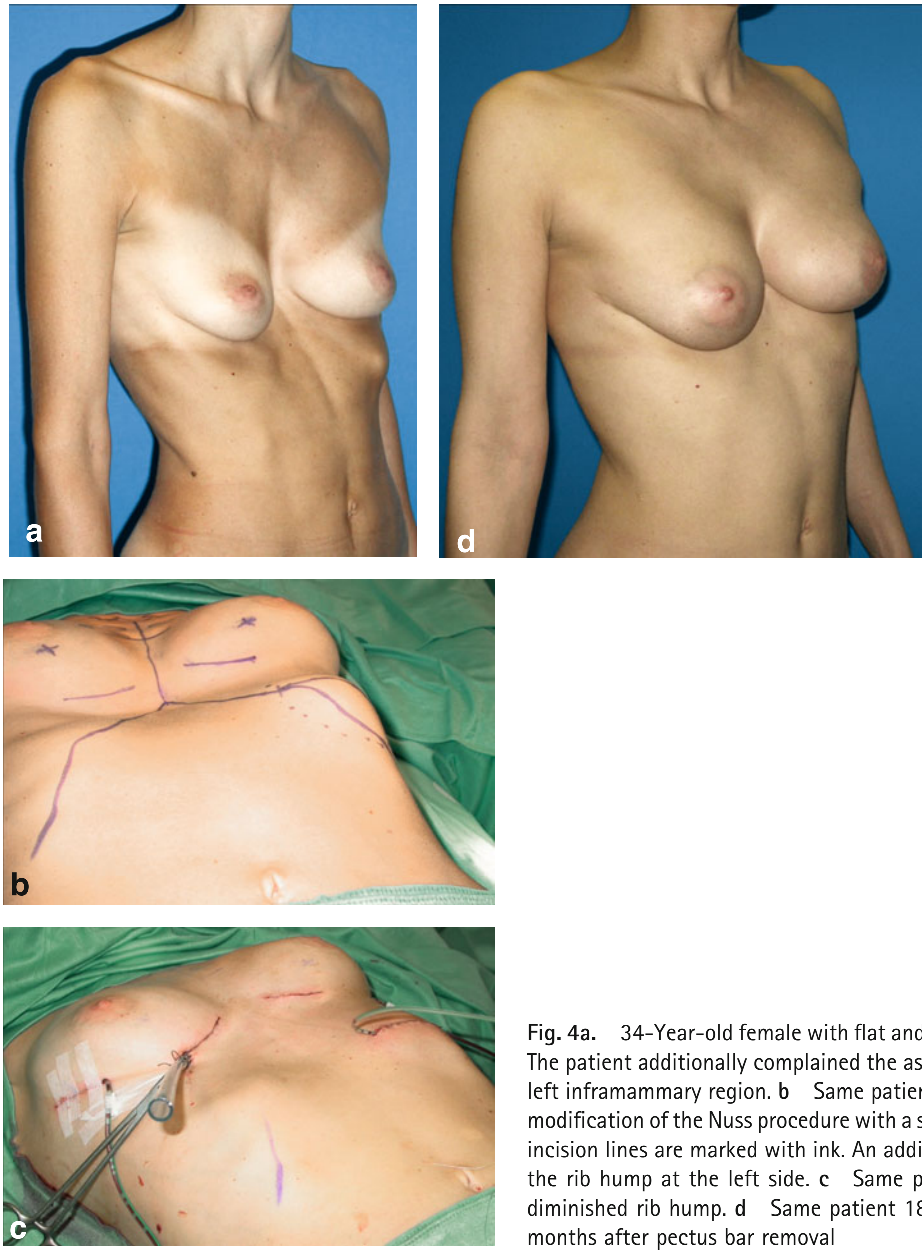

Fig. 4a. 34-Year-old female with flat and asymmetric funnel chest deformity. The patient additionally complained the asymmetric rib hump deformity at the left inframammary region. **b** Same patient intraoperatively, she underwent a modification of the Nuss procedure with a semi-open approach (MOVARPE), the incision lines are marked with ink. An additional incision is planned just above the rib hump at the left side. **c** Same patient after completed surgery and diminished rib hump. **d** Same patient 18 months after initial surgery and 6 months after pectus bar removal

Only in particular cases, and under certain circumstances, when a spontaneous regression may not be expected over time, a primary correction can be undertaken simultaneously to the pectus bar implantation already. This may be the case in elderly patients with maturated and rigid skeletal structures or in distinct asymmetric expression of such an associated deformity (Fig. 4).

Isolated rib humps may be also located parasternally or directly joining breasts in the submammary area in females (Fig. 6). They then can be corrected ideally by an incision along the submammary crease with the resulting scar hidden therein [11].

Isolated rib humps can appear also as a recidivation after surgical treatment of funnel, keel, or other [2, 7] chest deformities. They then correspond to isolated hypertrophic regenerates of still growing parasternal cartilages at childhood or youth. They almost never occur in bodily matured adults, but the probability is much higher when the patient is younger, particularly prior to growth spurts. If they are perceived as disturbing deformities by the patient, they may be corrected secondarily. In the funnel chest deformities, which are treated according to the MIRPE with conjoined chondrotomies or chondrectomies (MOVARPE), they may be corrected simultaneously with the explantation of the pectus bar. In recidivation of rib humps after keel chest correction (Fig. 7) as far as these are developed only at isolated levels, they can be resected even under local anesthesia, but

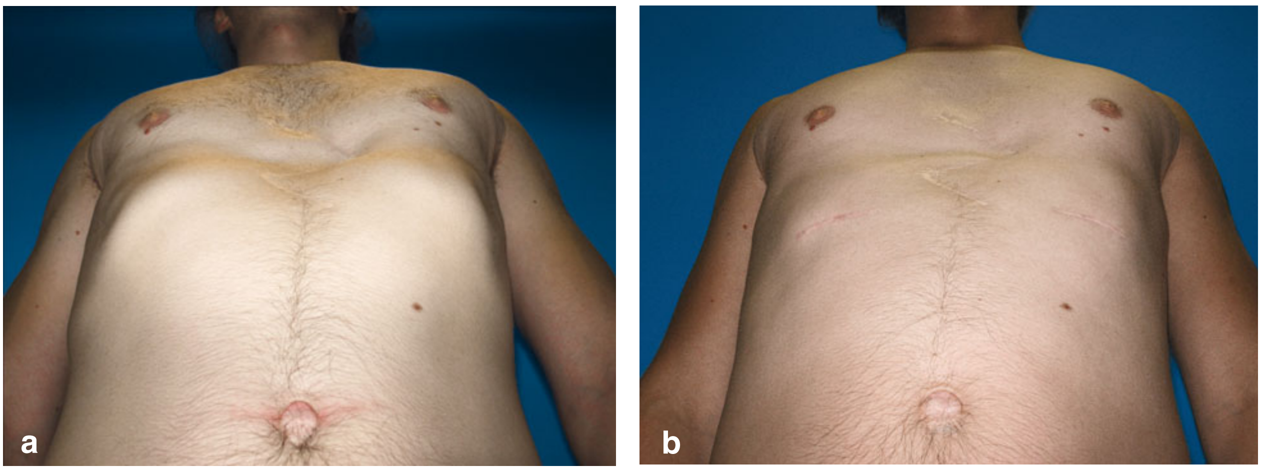

Fig. 5a. 28-Year-old male, 8 years after pectus excavatum correction using the microvascular sternum turn-over technique. Patient complained rib humps at the lower thoracic aperture and minor recidivation of the funnel depression. **b** Same patient 7 months after bilateral resection of prominent rib cartilages and bones. The cartilages were cut to chips and served as a subcutaneous augmentation of the residual funnel deformity presternally

Fig. 6a. 19-Year-old female with bilaterally developed rib humps, representing a minor form of keel chest deformity. Intraoperative situs depicting the planned incisions along the inframammary crease marked with ink. **b** Same patient 1 year after corrective surgery with minor visibility of the scars along the inframammary crease, only visible from beneath and arms elevated. **c** Same patient with well-hidden scars

Fig. 7. Isolated minor rib hump deformity at the xiphoid region parasternally left, resulting as a partial recidivation of rib cartilage growth, 1 year after pectus cartinatum surgery

not prior to scar maturation, which is first after 1 year, in order to prevent formation of over-excessive neo-cartilage.

Some authors apply videoendoscopically assisted thoracotomy (VATS, Chapter 5.4) for the resection of disfigured ribs from intrathoracally, in order to avoid external scars in the well visible area of the anterior thoracic wall [3, 6]. This means an advantageous, however skilful method preferably in females because the thoracoscopic port incisions are put far laterally, therefore result in very short scars only and furthermore lie far outside (of) the aesthetically concerned frontal aspect. The pertinent literature however predominantly deals with the treatment of rib hump deformities associated with scoliotic vertebral columns and fewer with minor deformities at the anterior thoracic area [1, 4, 5]. Nevertheless it means an outlook into a potential future development of a modern treatment of keel and funnel chest deformites.

11.1.1 Summary

The surgical correction of isolated rib hump or lower rib arch deformities is an easily performable intervention with regard to the minor invasiveness of such a surgical access. However, particular attention has to be paid on the protection of the intercostal nerves and pleura, furthermore on a careful surgical attention of the cut rib margins and in particular on the positioning of the skin incisions along the RSTL to gain an aesthetically pleasing result.

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11.2 Cleft sternum repair

Anton H. Schwabegger, Barbara Del Frari

Ectopia cordis because of its association with other congenital deformities and malformations as well as the nature of the deformity itself with instable cardiopulmonal function generally has a poor outcome with commonly unsuccessful attempts to surgical repair.

William W. Shaw [19]

As already described in Chapter 2.3.6 (classification) three groups of the expression of the sternum clefts exist which cause different therapeutic approaches [1–3, 14]. Because these sternum clefts, which occasionally present with ectopia cordis or as a Cantrell-pentalogy [4, 8] are subject of extremely rare deformities, accordingly the descriptions of surgical corrections herein are singular and available only as case reports or small series [17].

The indications for the correction of congenital sternum clefts are on the one hand restoration of the bony protection of the mediastinal structures and to restore normal intrathoracal pressure concerning breathing function, on the other hand, in wide clefts to prevent paradoxical movements of the intrathoracal organs. Last, and this above all only with less distinctive deformities, an aesthetic correction of the often well visible or palpable deformity (Figs. 1 and 2) is aimed.

In the literature a variety of different methods of the correction are described. They mostly deal with a bridging structure brought in to the defect, on the one hand with autologous cartilage. Such grafts can be transposed to the defect either as a pedicled or a free graft [12, 18]. Other authors [16] for instance transpose, that is turn-over the periosteum of the sternum bars to medial in order to cover the mediastinum as a first layer and then implant autologous cartilage grafts which subsequently are covered by advancement of pectoralis major muscle flaps. This method is suited very well when incomplete clefts are present and the width of the gap itself matches the size of periosteum flaps available from both

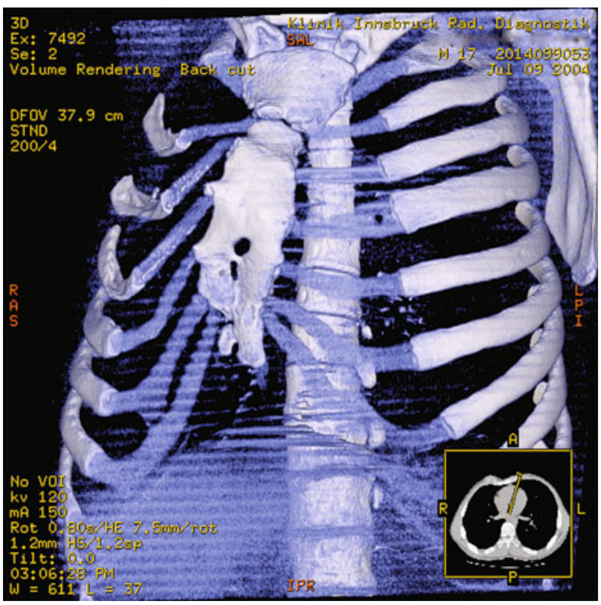

Fig. 1. Volume rendering 3D CT scan of an 18-year old male with major pectus arcuatum deformity and minor cleft sternum deformity

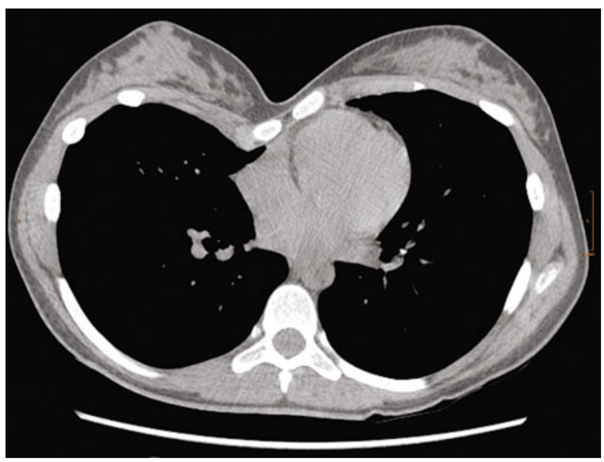

Fig. 2. A 16-year-old female with a sternum foramen associated with a pectus excavatum deformity, predominately complaining the funnel deformity but also the palpable bony deficit at the sternum midline

sides. Because, on this occasion, the intrathoracal diameter is not altered, the danger of intrathoracal compression barely exists with this method, even if mediastinal tissue, parts of the heart or the lung lie within the gap and can be repositioned without strain.
Other authors on the contrary [11] use titan plates, because this material provides with very good osseointegration and is well tolerated from the body, under circumstances needed to remain in situ for life.

Fig. 3. Newborn with thoracoabdominal ectopia cordis (Picture with courtesy of Prof. J. Hager MD, Department of Pediatric Surgery, Medical University Innsbruck.)

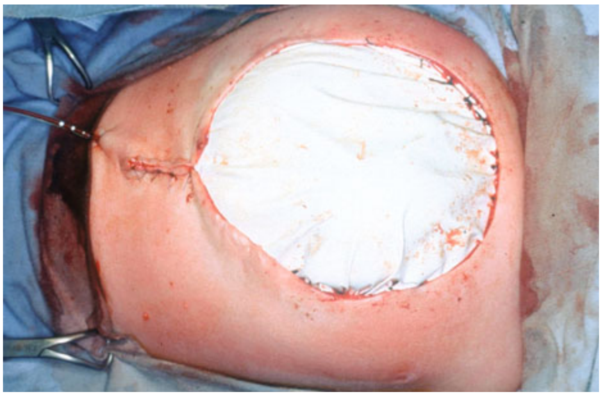

Fig. 4. Same patient as in Fig. 3, staged approach with sequential redressing of exteriorized organs by alloplastic material as a prearrangement for further defect closure. Despite absence of any compression but due to complex and severe cardiac malformations with subsequent cardiac failure the patient could not be salvaged (Picture with courtesy of Prof. J. Hager M.D., Department of Pediatric Surgery, Medical University Innsbruck.)

De Campos [5] and other authors [7] recommend the correction (primary repair) already in the neonatal period, if the patient considers in a stable cardiopulmonal state. At most the correction should be carried out as early as possible [6, 9]. As an adjunct to occasionally hazardous direct closure in ectopia cordis (Fig. 3), based on the risk of cardiac or major vessel compression, a temporary bridging of the defect with alloplastic material for staged redressing (Fig. 4) of ectopic organs into the thoracic cavity can support further surgery.

11.2.1 Discussion

Generally these methods of defect bridging should be preferred, which do not alter the intrathoracal diameter and would not result in organ compression sequel. That means, an approximation of the sternum bars should be avoided anyway, because otherwise the intrathoracal diameter narrows and can lead to compression and malfunction of heart and lung. To a certain extent the pressure can be compensated by flexibility of the diaphragm down to caudal but the limits are reached soon and may be recognized by reduced respiratory excursion, paradoxic breathing and lowering oxygen uptake.
Some authors also use allogenic material, for example Marlex® or Goretex® to bridge a defect [13]. Nevertheless, this goes along with the danger of infection and the inability of such inert material to adapt to the physical growth during childhood and later on. At most alloplastic material can be used to strengthen tissue sutures or for approximation of advanced Pectoralis major muscles [16].
The transection of parasternal rib cartilages for the purpose of an approximation of the sternum bars should be likewise avoided, because through this maneuver extensive and rigid scar formation might result and thus will lead to a severe growth obstacle till adult's age [10]. A severely constricted anterior wall with a still elongating vertebral column during growth spurt may inavoidably lead to a so-called hunchback deformity.
The direct defect closure during the neonatal period is absolutely the easiest way, because at this age the skeletal structures still show very softy and pliable features. The mobilization of the clavicles and the insertions of the sternocleidomastoid muscles however still remains a matter of controversies.
Suri in 1996 [21] reports about the correction of a complete congenital sternum cleft in an adult and

herewith used an iliac bone graft with VY-advancement of both pectoralis major muscles.
Sabiston [18] on the other hand used metamethacrylate muffed in Polypropylene to bridge sternum clefts. Above all in adults an approximation of the sternum bars can hardly be performed due to the rigidity of the thorax skeleton [21]. If herewith strong forces are employed, this maneuver will result in a reduction of the breath excursion and thus in a restriction of cardiopulmonal efficiency. Therefore, preferentially a reconstruction of the defect should be performed in adults, on the one hand either with autologous material [15, 19, 20, 22] or, on the other hand, even with alloplastic material like silicone, Goretex® or titan plates. Although these alloplastic materials can achieve a better stability compared with autologous tissue primarily, the latter is still to be preferred, because here less risk of infection exists and later reactions to alloplastic material is circumvented. Besides, later intrathoracal interventions in the area of the sternum are complicated by the presence of such alloplastic materials, in particular from metals.

11.2.2 Conclusion

It is recommended to correct sternum clefts already in early infancy, provided that they are recognized at birth yet. Such an early surgery is suggested because of the yet very elastic skeletal components of the thoracic wall with highly compliant ability to adapt cardiopulmonal function to altered biophysical conditions. The technique of approximation of the sternum bars is recommended only if a narrow gap between the sternum bar edges exists and such a minor approximation is estimated to cause no cardiopulmonal restriction. On the other hand, multiple parasternal chondrotomies should be avoided, in order not to impair the later growth of the anterior thoracic wall by scarring. In addition to that, more spacious defects must be bridged preferably by autologous cartilage grafts to circumvent restriction of the thoracic cavity that would otherwise result from extensive narrowing of a gap.
The experiences of the available literature during the past decades are based on single case series. It therefore may be assumed that in sternum clefts of minor extent above all in the neonatal period an approximation of the sternum bars is possible without restriction of lungs and heart function. At an advanced age in any case, as well as with wide clefts in the neonatal period, bridging procedures should be used. However, autologous tissue should be preferred in order not to restrict the growth capacity of the skeletal structures on the one hand, and to reduce the danger of infection in adults on the other hand.

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Complications, special problems

12

12.1 Complications, special problems, tips, and tricks

Anton H. Schwabegger

The recurrence of a thoracic wall deformity, months or years after shaping, can be avoided through no method with absolute security.

Surgery at the anterior thoracic wall can lead to specific problems peri- and also postoperatively because of the complex functional anatomy of the thorax and the complexity of the three-dimensional remodeling intervention. Above all the learning curve is relatively high-angled in the early stage of acquiring different surgical techniques of the individual procedures [27, 43]. Particularly and predominantly operation steps straightened on aesthetic improvements but lack of experience and lack of awareness of the functional anatomy let arise mistakes with more or less undesired or even drastic results for the affected patient. Severe complications were observed in the early period of applications, predominantly due to lack of experience. They usually subside with increasing numbers of patients and frequency of surgery within decreasing timeframes [24, 26, 30]. In this chapter, ample scope is dedicated for the subject of complications that may occur during and after the corrections of the funnel chest deformities and their variations, as well as the keel chest deformities. In specialized clinics, this kind of surgery dealing with congenital deformities of the anterior thoracic wall may represent a routine intervention with generally a poor risk incidence. In spite of the largest experience and specialized care, complications can nevertheless appear within all methods of surgery and postoperative treatment, eventually requiring acute or planned further interventions. Any treating physician must be prepared for such problems and must be able to solve them or immediately consult specialists from other departments experienced in specific incidents that may occur especially in the endoscopic treatment of pectus excavatum. Nevertheless an interdisciplinary approach in endoscopic techniques is always of superior advantage if minor experience therein is present, in order to provide with optimal surgical skill, technical experience, and the preparedness of specialized know-how onto potential complications.

Particular complications related to specific techniques are described in the relevant chapters also.

12.1.1 Peri- and postoperative as well as long-term complications in the correction of pectus excavatum deformity (in alphabetic order)

12.1.1.1 Adverse effect

In an unfavorable case with asymmetrical cartilage growth, this may even result in asymmetrical elevation with twisting of the sternum (Fig. 1) and subsequently to a cumbersome pectus arcuatum deformity. However, this carinatum or arcuatum deformity can be corrected surgically utilizing the conventional techniques of keel chest surgery, but it requires an additional intervention. There is no evidence or experience published on correction of such an iatrogenically triggered deformity, but from the viewpoint of cartilage growth this process

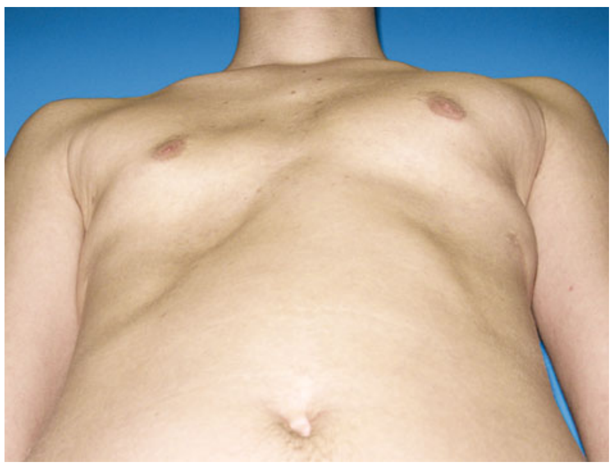

Fig. 1. A 23-year-old male with remaining asymmetry and some minor extent of reactive keel chest deformity at his left side due to incomplete derotation of a major malrotation. Pectus bars remained 3 years in situ; photo is taken the day prior to explantation

should be watched and temporized possibly for 1 year before corrective surgery is performed.

12.1.1.2 Allergy

Unknown allergy, first recognized after implantation of the metal hardware, can lead to fluid discharges at the implantation site, skin inflammation with rash along the skin overlying the bar [26], or low symptomatic pleural effusion. The main reason for such incompatibility with tissue is an allergic reaction against the Nickel component (up to 26%) in the metal struts or pectus bars. Major reactions may indicate the preliminary explantation of the pectus bar. In the case that

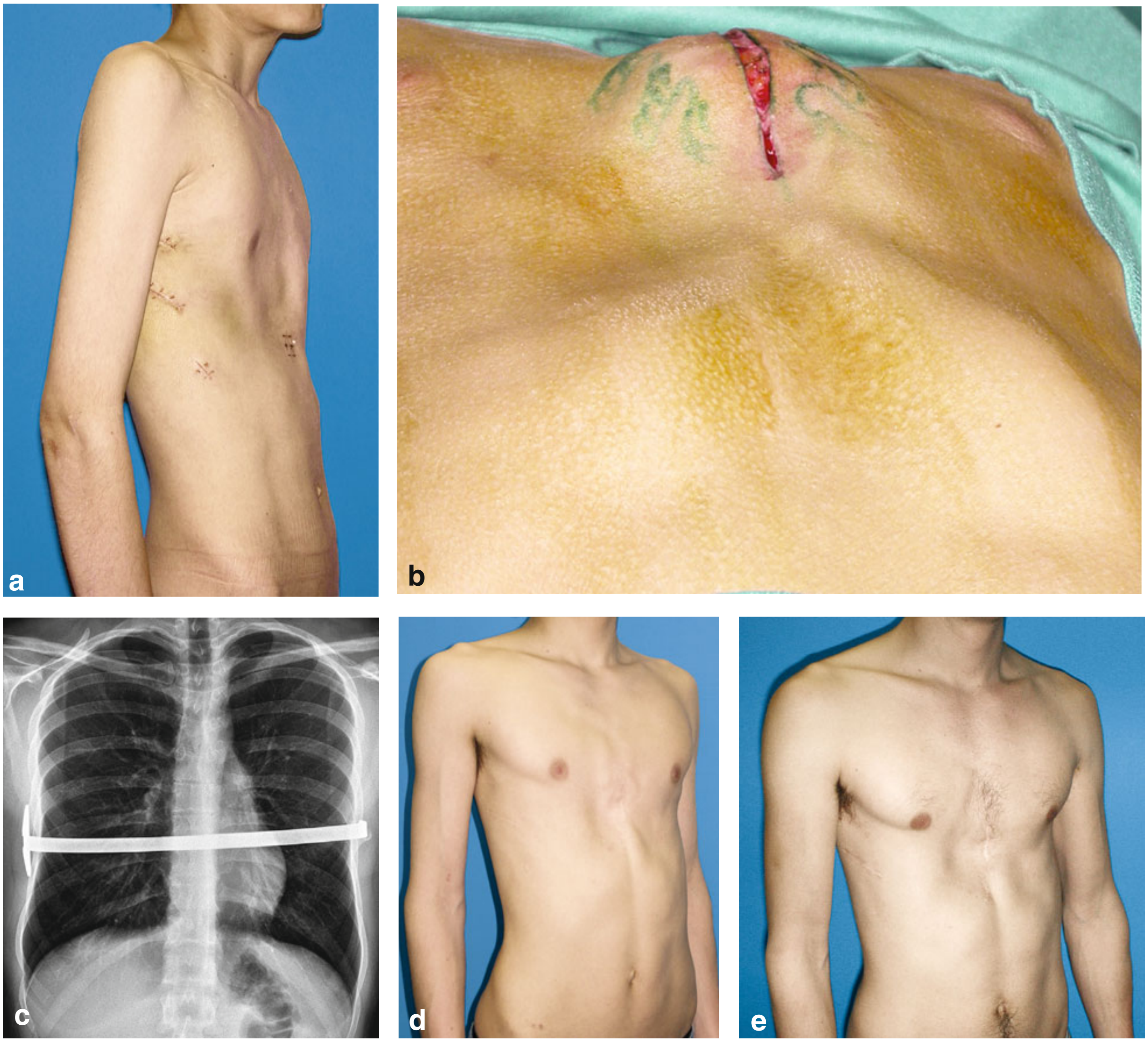

Fig. 2a. Mild reactive pectus carinatum parasternally left, 2 weeks after MIRPE with two pectus bars in a 14-year-old boy. **b** Intraoperative sites to correct a sudden onset of an increasing reactive keel chest deformity, same patient as in **a**. The deformity occurred within several days during growth spurt but first 8 months after initial surgery. **c** Too tight application of the pectus bar wing and fixed stabilizer to the thoracic wall in another patient, resulting in narrowing of the transversal thoracic diameter at the patient's left side during growth spurt. **d** Same patient as in **a**, 26 months after reactive keel chest correction by a conventional Ravitch procedure, 2 months of keel chest bracing and prior to pectus bar removal. **e** Late result in the same patient at the age of 20, 1 year after explanation of the pectus bars, which until then remained in situ for 5 years upon strong desire of the patient and fear from another relapse due to the reactive deformity years ago

Nickel allergy is well known preoperatively, alternatively a titanium strut or bar may be fabricated, custom made to the desired shape of the patient's thorax. However, such a preformed titanium-made strut is hardly malleable any more and therefore must perfectly fit prior to implantation. On the other hand, such preformed and thus not unbendable bars raise difficulties during explantation with the danger of intrathoracal organ lesions. In such cases and predominantly in minor deformities it might be advantageous to apply alternative surgical methods with low risk.

12.1.1.3 Acquired deformities

Immediately but more likely weeks and months after implantation, when a very elastic central part of the thoracic wall and preferably when a rather deep funnel depression with steep inclination is present, the pectus bar suspension can cause an adverse effect on the thoracic wall, namely the formation of an iatrogenic keel chest deformity (Fig. 2a and b) This reactive effect [8, 13, 39, 45] is more likely when deeper funnel is present and if during puberty the physical growth leads to an excessive growth of the length of the rib cartilages. In such a phase of growth spurt, if the lateral wings of the pectus bar limit the lateral expansion (Fig. 2b) of the skeletal thorax too early, rapid cartilage growth leads to elevation of the sternum and the adjacent ribs in the median line (Fig. 2a). Corrective surgery according to the conventional Ravitch procedure with postoperative keel chest bracing may finally lead to pleasing result (Fig. 2d and e).

Thoracic scoliosis or other constriction on the anterior thoracic wall may be caused by too early and especially too extensive surgical remodeling with subsequent scarring of a large area of the anterior thoracic wall [6, 14, 21, 25, 35]. Such interventions in early childhood and prior to any growth spurt with conventional remodeling techniques will inevitably lead to heavy scarring of ribs, sternum, and adjacent musculature necessary for breathing. Severe symptoms of respiratory distress [35] with exercise, primarily abdominal breathing, and failure of chest wall growth may result in the so-called acquired Jeune's syndrome [14]. Such extensive surgical interventions, if at all necessary in the early childhood already, should nowadays be replaced by the MIRPE method, which leaves most of the skeletal structures with adjacent soft tissue untouched.

Simultaneous (Fig. 3a) implantation of a custom-made silicon block together with a breast implant may yet correctly equalize an asymmetric thoracic wall incongruity (Fig. 3b) but at the same time will cause an unpleasant distortion of the female breast (Fig. 3c).

Pseudosymmastia may occur after simultaneous implantation of a custom-made funnel chest silicone block with breast augmentation implants. The wide subcutaneous undermining at the presternal region may not sufficiently separate the prepectoral pockets of mammary implants, thus resulting in a confluent large pocket. Herein the mammary implants unsteadily glide along the surface of the funnel implant and result in breast distortion medially, causing iatrogenic symmastia (Fig. 4a). Such deformities are extremely challenging and may require several surgical steps and ensuing long-lasting application of a compressive brace (Fig. 4b) to reach a sufficiently aesthetic outcome.

12.1.1.4 Arrosion, pathological rib fracture

Physical growth in youngsters or adolescents can lead to specific long-term complications after prolonged duration of implantation rest of a pectus bar. The lateral wings of the pectus bar progressively decrease their distance to and thus more narrowly join the bony ribs due to the growth of the rib cage.

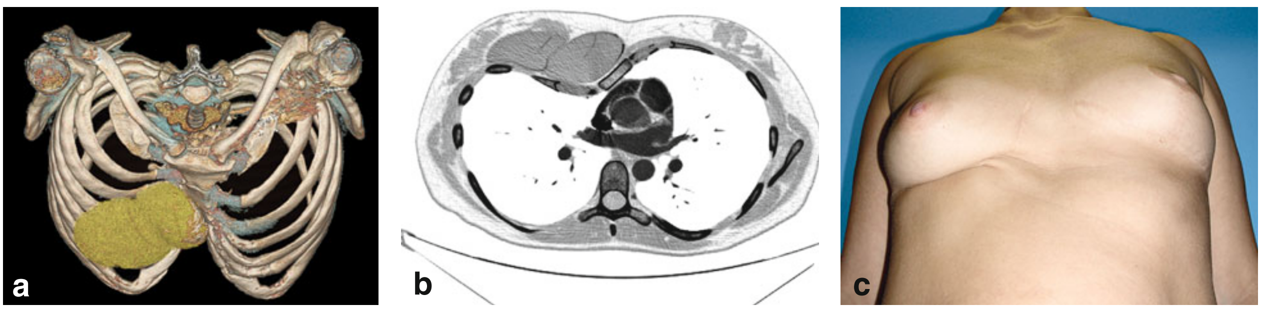

Fig. 3a. A 38-year-old female underwent correction of an asymmetric pectus excavatum deformity with simultaneous implantation of two custom-made silicone implants. **b** CT scan of the same patient, revealing sufficient augmentation of lacking chest volume by these implants. **c** Both implants with undue weight could not sufficiently be suspended by tissue and lead to unpleasant ptosis and distortion of the right breast

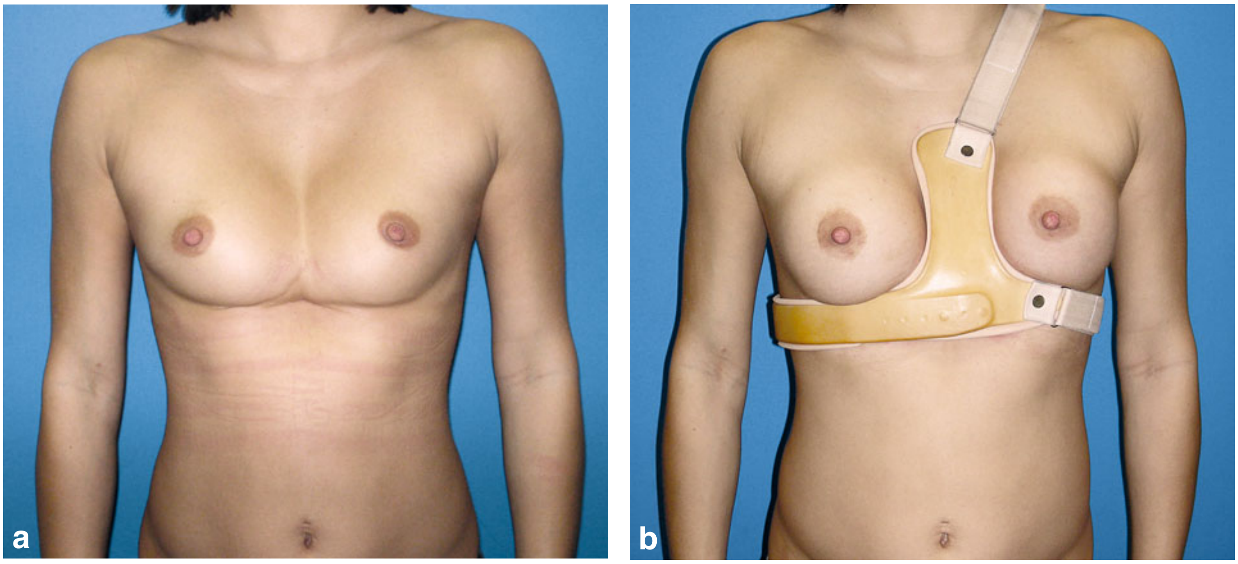

Fig. 4a. A 25-year-old female with a central custom-made funnel chest implant and bilateral breast implants, simultaneously inset and causing distorting pseudo-symmastia. **b** Removal of the central silicone block and surgical repositioning of the breast implants needed bracing for 3 months with moderately sufficient result thereafter

The permanent stimulus resulting from respiratory movements may lead to arrosions of the periosteum and bone with subsequent creation of an osteoclastic hole. Also primarily during implantation too narrow arrangement (Fig. 2b) of the lateral wings and/or stabilizer of the pectus bar to the ribs can cause these problems.

A complication resulting from such arrosions [23, 34] may be a pathological and painful rib or sternum fracture at the contact surface of metal hardware with the ribs. It may also happen after explantation of the metals, when the bony impression is weakened through removal of the implant itself or by resection of perifocal callus (Fig. 5a and b). This fracture due to weakening of the bone continuity can spontaneously appear just by change of position in the bed or by a cough and then lead to sudden violent pain. Apart from palpation, a conventional X-ray picture can localize the fracture, if this is not diagnostic a computer tomography is helpful. Usually the pectus bar is not to be removed but a cingulum (circular compressive bandage) may be helpful to diminish or release pain.

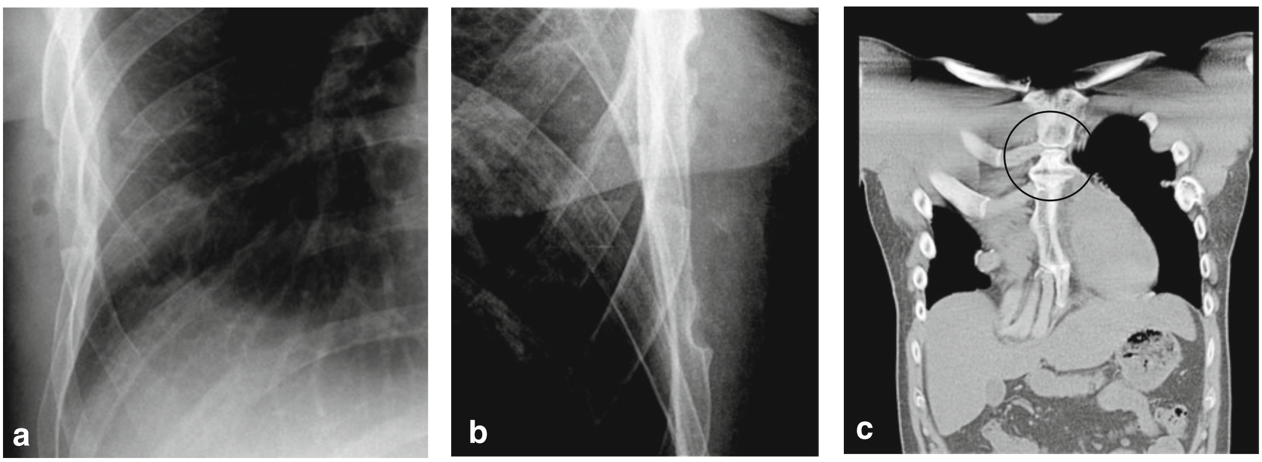

Fig. 5a and b. Arrosion of ribs with reduction of diameter thus weakening by too tight fitting of pectus bar wings and stabilizers. **c** Horizontal arrosion of the sternum at the 2nd intercostal space. The pectus bar tilted cranially with its central (median) edge riding on the posterior surface of the sternum bone, causing weakening, pain, and reactive callus formation by permanent friction (circle)

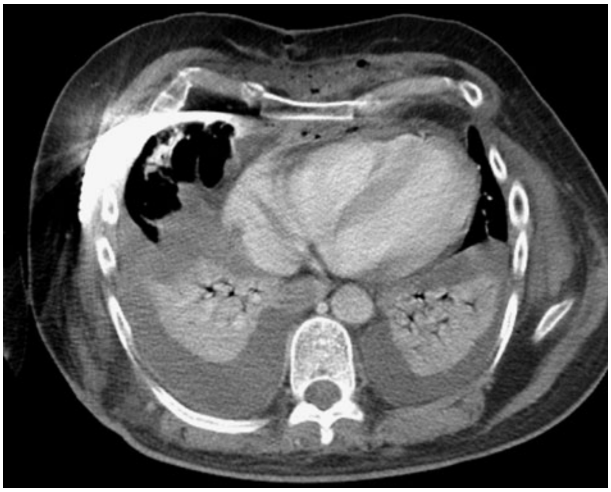

Fig. 6. Atelectasis at both basal lung lobes caused by hemorrhage and simultaneous pneumothorax in this 42-year-old male with a modified open repair after several corrective surgeries since childhood

High and continuous pressure of a pectus bar, usually present in aged adults, may even cause arrosion of the sternum bone (Fig. 5c) potentially resulting in pseudarthrosis and ensuing pain because of instability.

12.1.1.5 Atelectasis

Pulmonary atelectasis (Fig. 6) is mainly a problem in adults and is particularly caused by pneumothorax, major hemorrhage, or long-lasting pleural effusion with considerable amount of intrathoracal fluids. Usually such incidents are easily detectable by routine radiographic investigation during the postoperative stay in the hospital. Later on, after demission the symptoms such as dyspnea or pain at the back must alert the physician to perform clinical examination (percussion) and radiographic imaging. Symptomatic pneumothorax and hemorrhage need to be treated immediately by surgical exploration and intervention in order to keep the extent of atelectasis to a minimum. Minor pleural effusion should be routinely controlled and aspirated if increasing or symptomatic. Usually minor effusions resolve by time and with ongoing respiratory exercises.

12.1.1.6 Bleeding, hemorrhage

Stronger hemorrhage during surgery or within first few days after the intervention is a very rare event and transmission of blood or blood components can become only rarely required. Bleeding can appear by injuries of the intercostal vessels at the site of perforation of the

Fig. 7. Skeletal specimen depicting the flat introducer and potential complications by shearing effects on the subcostal neurovascular bundles as well as on the rib periosteum

pectus bar at the thoracic wall. Simultaneously with forced elevation maneuver at the sunken sternum, the perforation at the intercostal spaces and introduction or pulling through of the introducer and subsequently the pectus bar have to be managed. Therefore, the neurovascular intercostal bundles are prone to lesion (Fig. 7) and it is therefore required to inspect such perforating maneuver via endoscope (Fig. 8). That can securely guide the introduction and passage of instrument and pectus bar along the cranial margin of the rib circumventing damage of the inferiorly running intercostal vessels and nerve (Fig. 9).

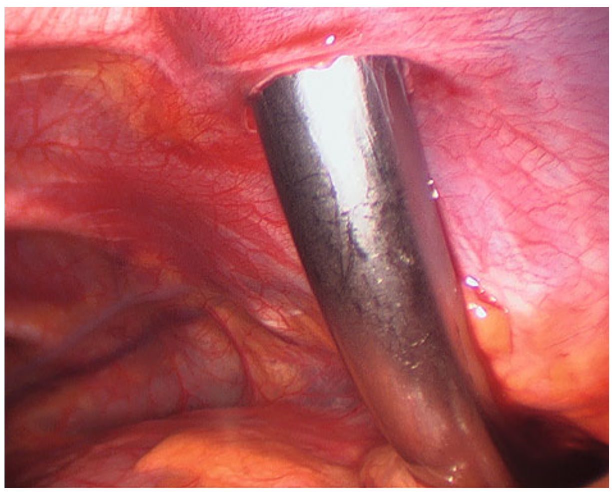

Fig. 8. Endoscopic view of the round introducer tunnelizing behind the sunken sternum, passing posterior to the transverse thoracic muscles which protect the internal thoracic vessels (oblique course at lower left edge of picture)

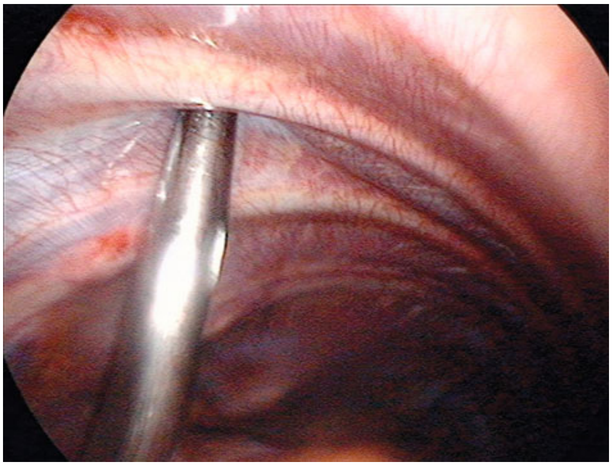

Fig. 9. Endoscopic view to the left thoracic cavity at the moment of perforation with the round tunnelizer. The intercostal neurovascular bundle may be well visualized and preserved at the lower rib margin (sulcus costae)

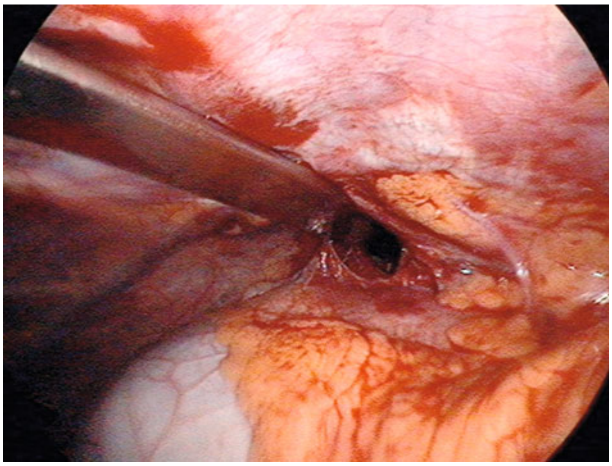

Fig. 10. Endoscopic view to the right thoracic cavity with the pectus bar in place, crossing the internal thoracic vessels (oblique course at lower left edge of picture)

A further source of bleeding may be due to the rupture of the internal mammary/thoracic vessel (Fig. 10) caused by forced introduction of the tunnelizer/introducer or the pectus bar. This complication appears more likely when a rigid skeletal thorax is developed, i.e. rather with the athletic male adult or with deep and steep funnel deformity [38, 43]. In these cases, increased forces must be applied to elevate the anterior thoracic wall, which is partially managed by the introducer and then subsequently by the flipping maneuver of the pectus bar. Due to the skeletal counterpressure the edges of the instruments then can lead to a shearing-off effect and squeezing off rupture of the internal thoracic vessels with

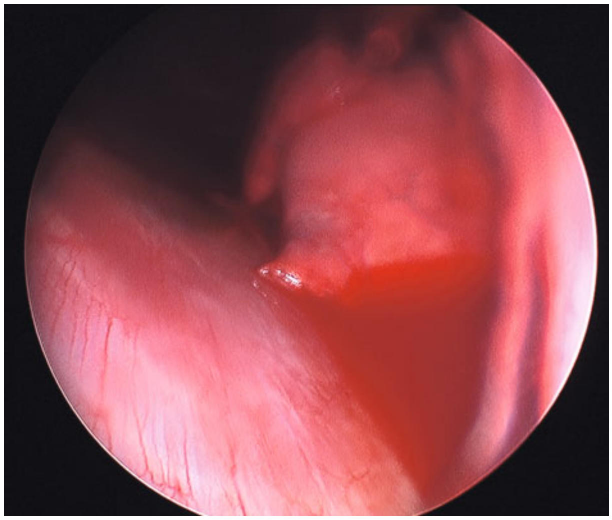

Fig. 11. Intrathoracic hemorrhage caused either by damage of intercostal or by internal thoracic vessels. The collapsed lung lobe is seen at the right side surrounded by blood

critical (Fig. 11) or even life-threatening delayed bleeding. Under such circumstances, false manipulations during the implantation maneuver or potential bleeding can be recognized on time only under simultaneous visualization (VATS, Chapters 5.4 and 6.3). For these reasons the utilization of thoracic endoscopy is indispensable with the MIRPE or a similar technique to protect not only the intrathoracal organs against fatal injuries but also the thoracic wall vessels. In addition, especially in adolescents and adults [38] with the need for increased manipulation forces it is recommended to use a rounded introducer/tunnelizer (Fig. 12) to avoid

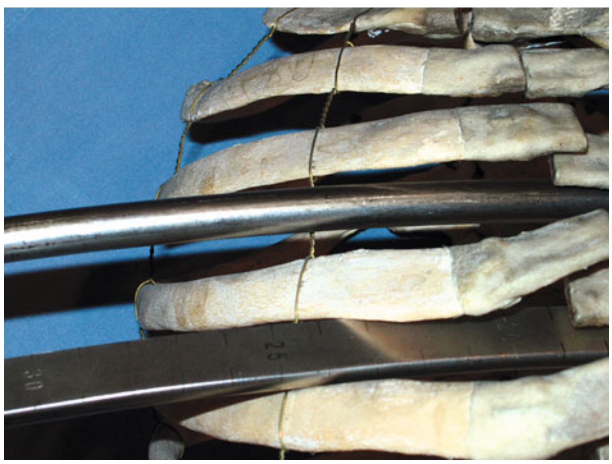

Fig. 12. Anatomical specimen depicting the relatively sharp edges of a strap tunnelizer versus the round one, introduced in separate intercostal spaces, to depict the higher risk of damage to soft tissue entrapment by the strap tunnelizer

edging and cutting of intercostal or internal thoracic vessels with immediate (Chapter 13.2) or late onset of bleeding from eroding effects on the internal thoracic vessels [1].

A search of the literature revealed only singular published cases of cardiac perforation or migration to the abdominal cavity during the MIRPE procedure, with no lethal consequences [4, 5, 12, 22, 24, 28, 40]. Liver piercing [5] may be a consequence of a too far caudally placed endoscopic port at the right side, which guides the endoscope to the intraabdominal space below the diaphragma. Generally the use of an endoscope (VATS) may circumvent inadvertent lesions of the thoracic organs by indirect but nowadays by high-resolution visualization (Chapters 5.4 and 6.3).

In analogy to the use of a so-called Erlangen-Tensiometer [45] for the measurement of the forces to be overcome during mobilization, that is elevation of the thoracic wall, the thoracic wall can be elevated by a simple bone hook (Fig. 13) intraoperatively, right before the tunnelizer is introduced and the pectus bar is implanted. This maneuver causes a flattening of the funnel to ventral, so that by the introduction of the tunnelizer and subsequently the preshaped pectus bar the passage obstacle behind the otherwise depressed sternum is alleviated. Thereby the danger of penetration of lung or pericard by misdirected insertion is distinctly minimized, apart from a much better survey and augmented manipulation space intrathoracally. Additionally hereby the forces to be overcome by insertion of the metal parts are reduced and with it the danger of injuries of the rib periosteum, the subcostal neurovascular fascicles, and also the retrosternally running internal mammary/thoracic vessels.

However, this elevation maneuver shows even other advantages. A special advantage consists in the fact that, on this occasion, the deformity is virtually preshaped and the remaining pressure on the implanted pectus bar is reduced from the beginning of remodeling. On this occasion, it can be estimated whether a second pectus bar is necessary for proper remodeling of the thoracic wall. For a quantitative determination of the forces hereby the Erlangen-Tensiometer [45] can be of additional value. Furthermore applying a minor-open pectus excavatum correction in combination with the pectus bar or other metal strut, this elevation maneuver permits a retrosternal mobilization, and under circumstances it also enables a necessary separation of intrathoracic structures from the sternum.

Such an elevation maneuver to improve the intrathoracic survey may elegantly also be performed utilizing the vacuum bell after Klobe (Chapter 6.8), either as an adjunct measurement preoperatively or on the other hand simultaneously during surgery (Figs. 10 and 11 in Chapter 6.8).

Fig. 13. A bone hook placed behind the lower part of the sternum for its elevation will enable or alleviate the passage of instruments through the thoracic cavity

Fig. 14. Subcutaneous hematoma at the postoperative day after implantation of a funnel chest silicone block, requiring immediate evacuation to avoid potential skin necrosis

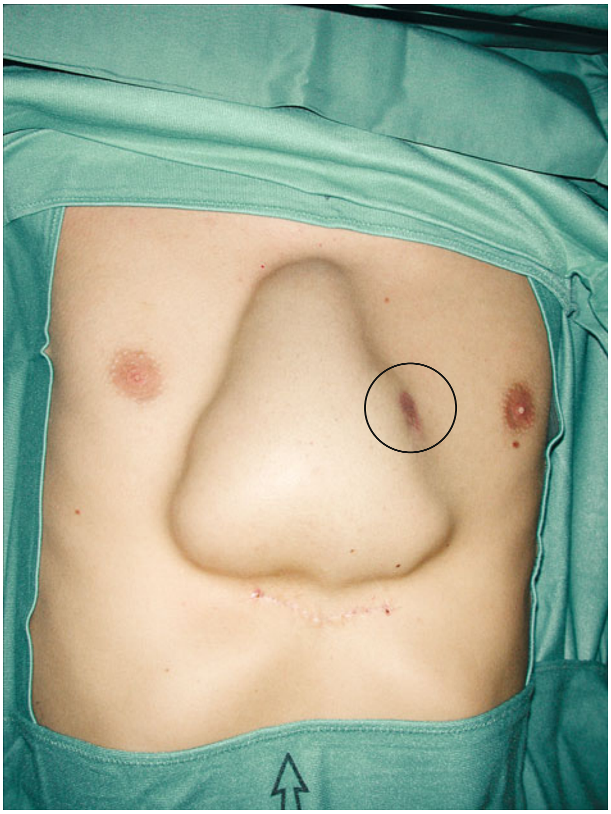

Fig. 15. Dangerous overdistension of skin by seroma formation with already beginning perfusion problems (circle) at the margin of the deformity at the patient's left side. Immediate revision is mandatory to avoid disastrous skin necrosis

Hematoma after wide subcutaneous dissection of a pocket to implant a custom-made silicone block is a common complication (Fig. 14) and must be treated as soon as detected by aspiration or surgical evacuation. Such an overdistension of skin caused by the volume of the hematoma may cause skin necrosis, as the presternal skin relies on random perfusion only after cauterization of parasternal perforator vessels to enable pocket formation. Likewise the occurrence of major seroma has to be treated on time due to the same reasons (Fig. 15).

12.1.1.7 Brachial plexus lesion

Injuries to the plexus brachialis are not related to endoscopic pectus excavatum surgery itself, but may be the result of overdistension during positioning of the patient. Over-elevation or unintended retroversion of the arms prior to surgery and lasting during surgery may result in temporary but in severe cases also in permanent nerve lesions with numbness or even muscular weakness and palsy [11]. Especially in the MIRPE or MOVARPE technique, the range of the dissecting instruments and endoscope depends on the proper positioning of the patient on the table (Chapter 5.2). Forced manipulation may loosen the arm supporting struts thus leading to unrecognized dislocation of the arms further to cranial or even dorsal with threatening tension on the brachial plexus.

On the other hand, if the conventional Ravitch technique or a similar method for the pectus excavatum repair is applied with extensive pectoralis major muscle elevation, the pectoralis nerve branches are prone to lesion. Emerging from the brachial plexus and initially running at the posterior surface of the muscle they may be damaged either by direct surgical trauma and overdistension from surgical hooks or by diathermic hemostasis. Such isolated nerve lesions may result in segmental muscle palsy with inadvertent aesthetically unpleasant appearance by muscle atrophy, which especially in athletic males is an eye-catching stain.

12.1.1.8 Callus formation

Such bony overgrowth occurs usually at the pectus bar wings, where they are fixed to the ribs laterally. Callus may also be present at the entry site of the pectus bar into the thoracic cavity, where it crosses the intercostal space. Shearing-off effects of the periosteum from the ribs herein via reactive periostitis cause the formation of callus primarily due to deperiostation by the introducer or the pectus bar during forced implantation maneuvers.

On the one hand, callus formation has the advantage that the pectus bar is fixed in such position that it cannot be dislocated any more; on the other hand, this callus can overgrow the pectus bar wings completely and therefore complicates its explantation (Fig. 16a–c). Excess callus, if well palpable, should be removed at the time of the pectus bar explantation procedure.

Palpable and under circumstances well visible callus may also grow after months at the site of a horizontally placed sternum osteotomy, occasionally required in particular remodeling procedures. This is more likely when osteotomy is performed through the whole thickness of the sternum bone and subsequently is not secured by osteosynthetic material, allowing excursions during respiratory movements and therefore stimulates a reparative osteoplastic process. Therefore and especially in cases with necessary complete transsection of bone exemplarily for the correction of sternum malrotation, the osteosynthesis requires increased attention.

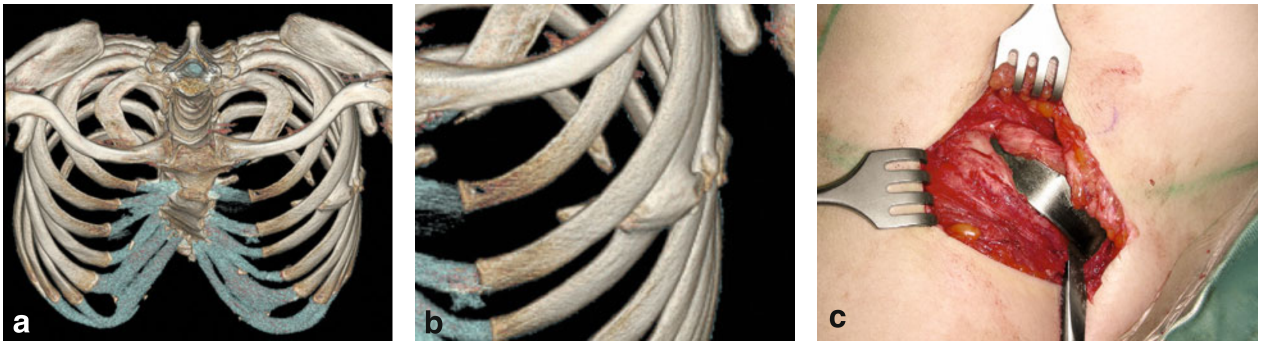

Fig. 16a–c. Callus formation at the entry site of the pectus bar. This complication is seen more frequent in adults due to the rigidity in contrast to the flexibility of ribs in children

12.1.1.9 Dislocation/shifting of pectus bar

Bar displacement and shifting (predominantly) to cranial also seem to occur more frequently and up to 9% [16, 26], when no lateral stabilizers are used in the MIRPE technique, or when no lateral and paramedian fixation sutures [17, 41] close to the sternum can be placed.

At the same time it has to be taken into account, however that the stabilizers have to be seamed to the surrounding tissue, or should be fixed with other fixation materials (e.g. riveting joint) to the bar in such a manner that gliding off is prevented. However, lateral stabilizers, if not fixed with circumcostal sutures and only to the soft tissue, will not sufficiently withstand the strength of the lateralized support of a rigid thorax.

The very strong pressure [9, 45] of the elevated sterno-costal unit, if the sites of perforation of the pectus bars were placed at the thoracic wall very laterally, may cause a dislocation of the pectus bar through disrupted intercostal muscles to the back. By such a sloping course directed along the ascending ribs to dorsal, the dislocated pectus bar will be pressed more cranially and, more cumbersome, into the thoracic cavity (Fig. 17). This dislodgement of the pectus bar to dorsal within the sagittal axis on the one hand causes that the sternum erection by suspension cannot be maintained any more and inevitably it will result in a relapse. On the other hand, this sloping of the pectus bar to cranial and dorsal, without real tilting or rotation is probably the most frequent complication and cause for the recidivation of a funnel chest deformity within adolescents and adults [5]. Such slipping may also occur along a longitudinal axis to cranial, the pectus bar approaching the site of horizontal osteotomy. In these cases forces are transferred adversely, causing an elevation of the mobilized sternum close to the osteotomy site but not at the area of interest. This adverse tilting of sternum (Fig. 18) will probably lead to pseudarthrosis at the osteotomy site with a development of a bony ridge that may be well palpable and visible. In children these problems are hardly given because of the substantially yet more elastic thorax and absence of distinctive rigidity. A too far laterally placed perforation port at the anterior thoracic wall for the introducer and the pectus bar as well as neglecting the distinctive pressure ratios within the athletic or matured thoracic skeleton are probably the most frequent initial mistakes of the utilization of the MIRPE technique, particularly when applied in adolescents and adults. It is of utmost importance to place the entry and exit points exactly at or slightly medially to the rim of the funnel, in order to transfer the forces of the pectus bar to the obliquely orientated ribs and not to the intercostal muscles [5]. Therefore, and above all in the adult patient or adolescents with athletic disposition as well as in defor-

Fig. 17. Pectus bar dislocated into the thoracic cavity by undue pressure after using the MIRPE with only one pectus bar in a 31-year-old tall male

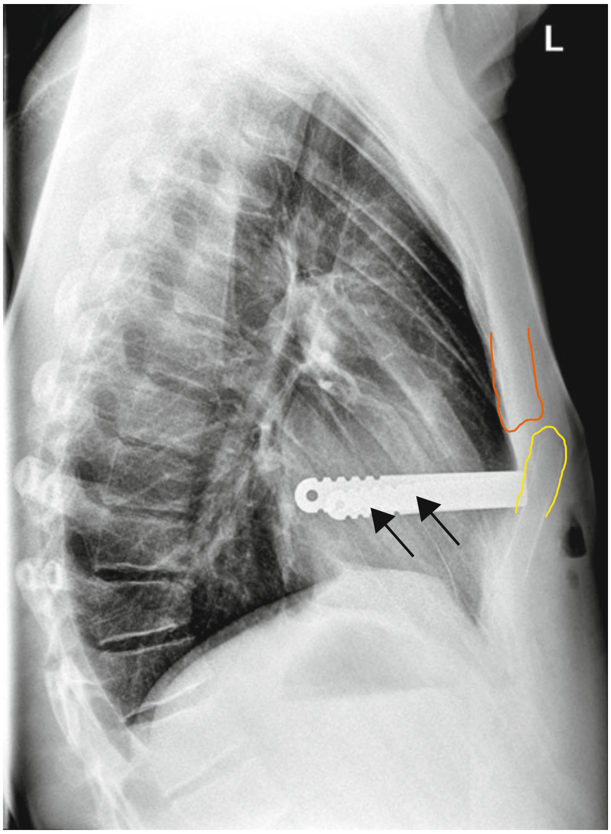

Fig. 18. Modified approach with MIRPE combined with horizontal osteotomy in a 196 cm tall and 16-year-old teenager. Dislodgement of the pectus bar (arrows), still placed in the right angle, but slipped too far cranially, causing adverse anterior tilting and pseudarthrosis of the sternum

mities with deep funnel and severe sternum inclination alternatively a combined approach (MOVARPE, Chapter 6.4) consisting of the MIRPE technique but with osteotomy and chondrotomies should be taken into account [9]. Not only acute and undue pressure load but also long-lasting pressure may cause a slipping of the pectus bar to dorsal, resulting in good visibility and palpability of the pectus bar wings. If such slipping, however to a tolerable extent only, is not avoidable because of the stature of a patient with a rigid skeleton, the use of shorter pectus bars is recommended in order to avoid dorsally placed bar wings. Such dislocation to dorsal may encumber the removal of the bar via the primarily set incisions thus requiring unsightly elongation of the scars. Even the pure strain load of a rigid and athletic thorax may cause splaying of the pectus bar, resulting in projected bar wings, very well palpable and visible then (Fig. 19), occasionally causing friction and irritation of the medial surface of the upper arm.

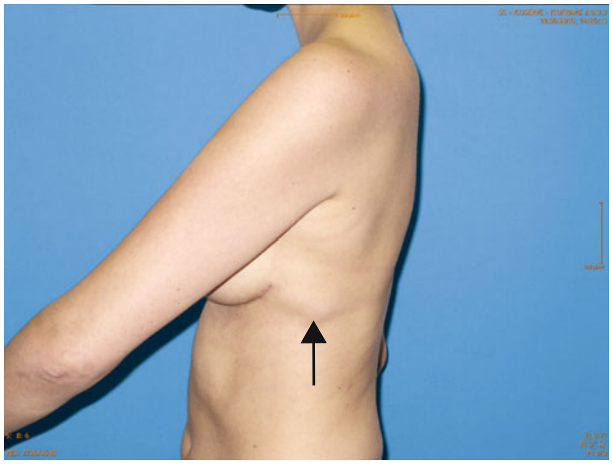

Fig. 19. A 24-year-old and 186 cm tall female with marfanoid features. Pectus bar wing splayed from the lateral thoracic wall due to the load of the anterior thoracic wall, despite osteotomy at the sternum and multiple parasternal chondrotomies. The projecting bar-end is well visible posterior to the breast (arrow)

12.1.1.10 Emphysema

Immediately after surgery present minor subcutaneous emphysema commonly requires no specific treatment and usually will disappear after a few days. If it is occurring later and is increasing, a CT scan is recommended to exclude a pneumothorax or a lesion at the lung lobes.

12.1.1.11 Floating sternum, pseudarthrosis

A floating sternum [32] may occur early or as a late complication many years after the initial treatment. It is complicated by symptoms such as significant sternal pain, instability of the anterior chest wall, and respiratory dysfunction. It may be caused by improper healing of an osteotomized sternum and failure of proper regrowth of partially resected costal cartilages (Fig. 20a and b).

12.1.1.12 Infection

Local infection at the site of implantation is a very rare event. It may be present when undue forces are applied to the incision margins with development of marginal necrosis with creeping infection into deep tissue layers. The use of lateral stabilizers however increases the incidence of wound trouble due to such tension maneuvers to skin, subcutaneous tissue, and muscles for placement of additional hardware [46].
A superficially infected bar wing must not be immediately removed as long as no systemic illness occurs [42, 46]. Local antiseptic treatment after surgical de-

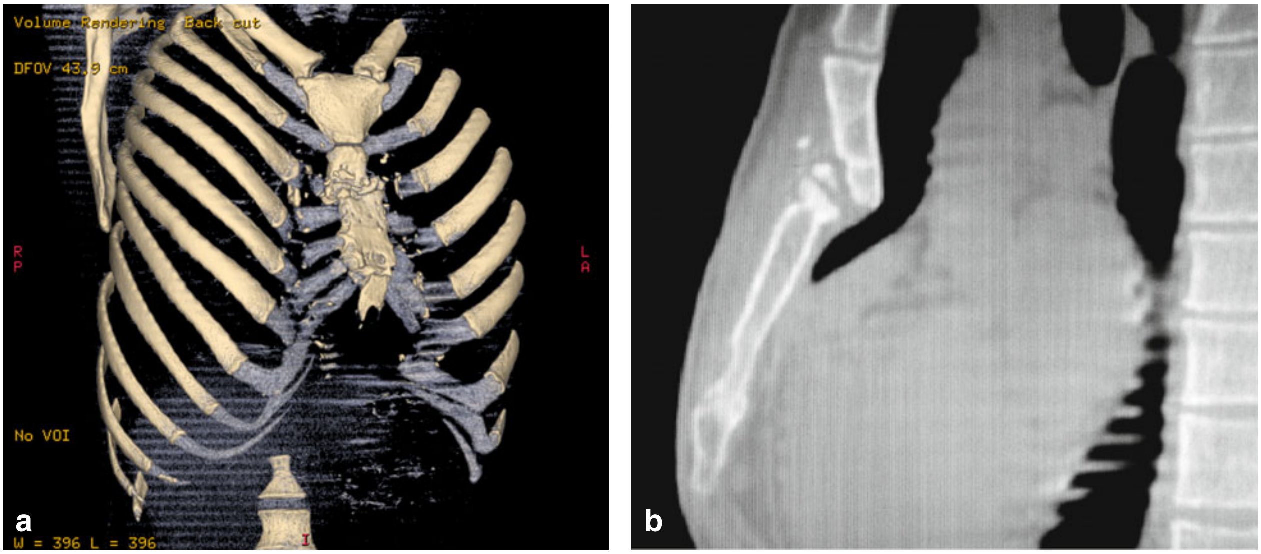

Fig. 20a and b. Pseudarthrosis at sternum at the level of the 3rd intercostal space after a microvascular sternum turn-over procedure, requiring revision due to permanent pain and crepitation sensation

Fig. 21. Chest tubes placed along a subcutaneous tunnel to prevent creeping infection into the thorax

bridement usually leads to healing. The ports of chest tubes need thorough care to avoid ascending infection. Their course may be placed through a long subcutaneous tunnel (Fig. 21) distally to the surgical access to minimize the risk of creeping infection along the tubes to the intrathoracal space.

Osteomyelitis of ribs or sternum is not a common problem within the pectus-remodeling surgery.

12.1.1.13 Mediastinitis

Isolated mediastinitis is also a very rare incident in performing thoracoplasties. Chest tubes, antibiotics, and intensive medical treatment might be necessary then upon onset. For many years this complication was very rarely seen and rather in the open techniques but may be a sequel of long-lasting or extensive wound infection at the pectus bar wings [44].

12.1.1.14 Neuroma, damage of intercostal nerve

Nerve injuries occur during every operation, as fine nerve branches will be cut in the subcutaneous layers. The consequence is a long-lasting or remaining feeling of numbness or reduced pain and contact sensitivity in the environment of the scar (Fig. 22), rarely perceived as disturbing.

Pressure damage at intercostal nerves on the other hand is a result of the forced implantation of the introducer or pectus bar. Pains rise immediately or within less than a week and leave behind usually only rare continuous troubles. More rarely formation of neuromas of intercostal nerves however may become a long-term sequela with parasternal or lateral thoracic continuously lasting pains. Such neuromas can hardly be localized and treatment with any method is often without success and therefore prevention of such iatrogenic nerve damage is of paramount importance. Such sequelae also must be contents of an informed consent as they may postpone an aesthetically optimal result behind the reduced quality of life due to permanent suffering from pain.

12.1.1.15 Pains

Wound pains should exist at most only immediately after the operation. Shoulder pain will resolve after days and

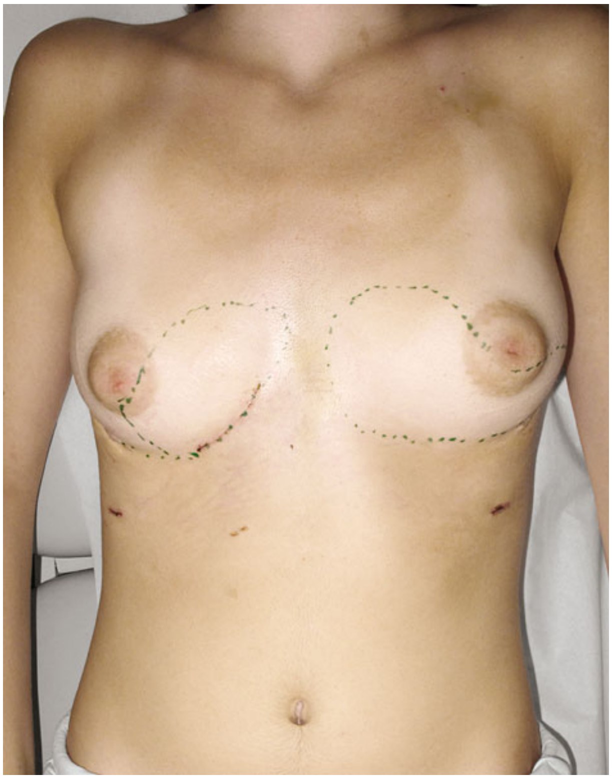

Fig. 22. Area of numbness at the lower breast quadrants due to irritation or damage of intercostal nerves either by the introducer, the pectus bar itself or by the circumcostal fixation sutures

usually is caused by the positioning in the MIRPE or similar techniques with required lateral access and elevated arms during surgery. Move pains and muscle pains in the weeks after the operation require no special treatment as they also resolve by time. Chest pain after remodeling surgery of keel chest deformity is kept low by peridural catheter and ongoing oral analgesics, furthermore by immobilization of respiration movements by the keel chest brace.

Longer lasting pain paravertebrally and at the back is caused by distortion of the costovertebral joints out of their original position due to the elevation of the sternum and the concomitant elevation of the ribs in the pectus bar techniques. It can be resolved by gymnastics and ventilation exercises.

Uncommon in children, but present in many cases of adolescents and even more in aged adults, the pure MIRPE technique with one or two supporting pectus bars will cause forced pressure against the back side of the sternum with ensuing long-lasting pain, frequently with the need of permanent intake of analgesics over months. This phenomenon may be circumvented in choosing a modified approach with sternum osteotomy and chondrotomies to loosen the rigidity of the anterior thoracic wall, thus reducing undue pressure against the sternum. In adults, the analgesic requirement is generally higher than in children.

12.1.1.16 Periostitis

See callus.

12.1.1.17 Pleuritis, pneumonia, pleural effusion, pericarditis

Such irritations rather than infections appear more frequently in sternum turn-over and other invasive techniques, also in MIRPE [24, 26] but rather more seldom in the Ravitch procedures [10]. The cause for effusion (Fig. 23) may be attributed to mechanical irritation of the pericardium and pleura during insertion. Later on such lesions may result from the edges of the metal strut itself, leading to permanent friction at the lungs and pericardium, especially and more, if the pectus bar is tilted and causes prominence into the thoracic cavity (Fig. 24a and b).

Infections following surgery of thoracic wall deformities are a very rare event [6, 26] although the implantation of foreign materials and above all iatrogenic and hardly avoidable injury of the pleura let us suspect a high rate of infections. The preoperative administration of single shot antibiotics has proven a positive

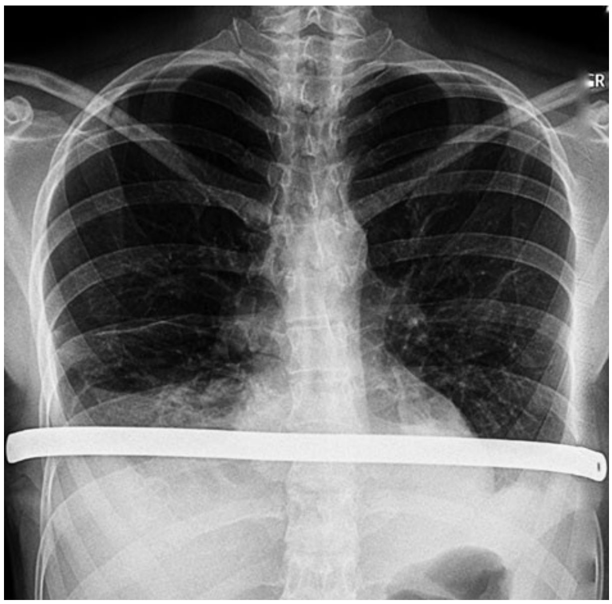

Fig. 23. Pleural effusion predominantly at the left thorax, with that amount and due to respiration impairment requiring aspiration or implantation of a catheter for drainage and to avoid atelectasis

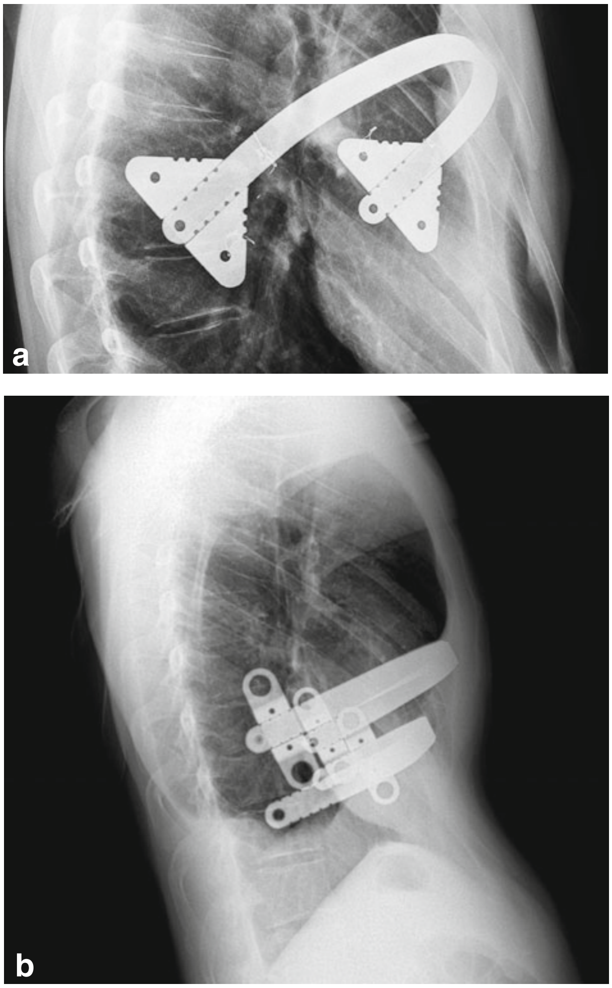

Fig. 24a and b. Tilted pectus bars with edging effects to the intrathoracal organs, causing pleural effusion and furthermore scarring with adhesions

Fig. 25. Rare case of pneumonia occurring at the second postoperative day after the MIRPE procedure in a young adult with increasing respiratory distress, requiring immediate removal of the pectus bar

effect in our patient's collective and according to the medical knowledge it is state of the art nowadays. Even with the temporary implantation of drainage tubes, the administration of single shot antibiotics should be sufficient along with suitable hygiene treatment at the drainage orifices at the skin. Occasionally but preferably in extensive open or minor-open techniques with particular circumstances like sternum osteotomy and extensive cartilage resections, the proceeding with antibiotic administration for several [3–5] days is recommended to prevent pneumonia (Fig. 25) or other infections.

12.1.1.18 Pneumothorax

A pneumothorax (Fig. 26a and b) is the corollary of the perforation of the thoracic wall with the implantation of the pectus bar. The extent of pneumothorax can be held low by raised ventilation pressure (PEEP) of the lungs by the anesthetist at the end of the operation but prior to the wound closure. A sickle-shaped pneumothorax without functional symptoms is detectable in about 50 or more percentage of patients treated with the MIRPE [26] and usually will undergo spontaneous resorption within few days. A routine implantation of chest tubes therefore seems not be necessary [31]. However, the implantation of chest drains to prevent development of pneumothorax is recommended in adults for several days under certain circumstances, such as higher age or athletic habitus with more skeletal rigidity, intensive surgical manipulation, or minor-open technique (MOVARPE). Schaarschmidt et al. [36] has succeeded it in a large series in creating an intrathoracal but still epipleural access with enough space for the pectus bar manipulations by endoscopic dissection, however, therefore in avoiding the sequelae of a pneumothorax. Another advantage of this ingenious however technically skillful thus more demanding and time-consuming method also seems to be the reduction of the infection risk (pleuritis or pneumonia).

A further potential cause for the origin of a pneumothorax is circumcostal fixation of the pectus bar or the stabilizers to the underlying ribs with sutures or

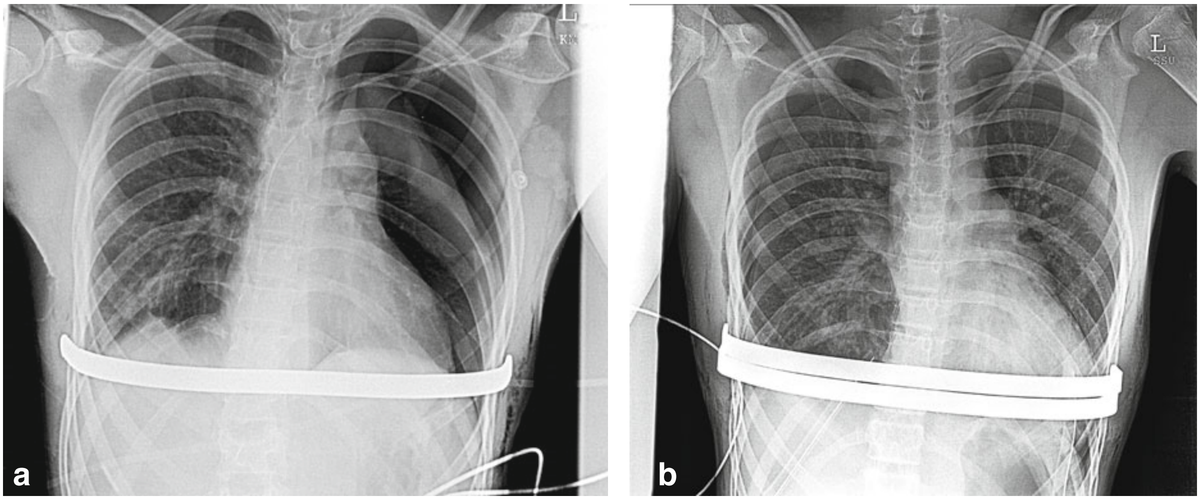

Fig. 26a. Massive pneumothorax at the left side with the urgent need for evacuation via chest tube. **b** Pneumothorax with moderate extent in a 16-year-old male, but due to bilateral presence and respiratory impairment required implantation of chest tubes

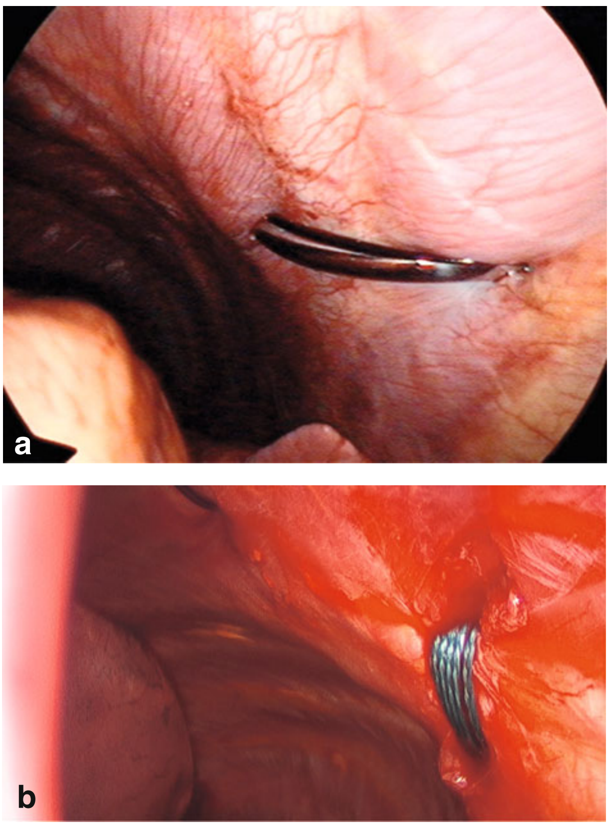

Fig. 27a. PDS sutures inserted using a Deschamps needle under endoscopic visualization, depicting a however minimal lesion of the parietal pleura. **b** Non-absorbable thread placed around one rib to prevent the pectus bar wing from tilting. Such minimal lesions to the pleura may also lead to development of pneumothorax

threads (Fig. 27a and b), which commonly are utilized to prevent tilting of the pectus bar out of the horizontal plane. With such sutures around the rib, may it be laterally or parasternally, the risk of a pleural rupture always exists. Particularly, laterally a laceration of the parietal pleura closely adjoining to the thoracic wall can hardly be avoided and a multi-layered watertight wound closure is mandatory in order to prevent invading of air and subsequent pneumothorax with collapsed lung.

Another eventuality for the generation of a pneumothorax originates during the removal of the chest tubes. Because of remaining in situ for several days and the utilization of mostly large-caliber drainage tubes, the growing granulation tissue all around the tube surface creates a rather fixed tissue duct. During extraction of the drainage tubes inadvertent respiration movements can lead to aspiration of air along this tissue duct. Therefore, it is advisable to place the site of skin perforation of the drainage through subcutis and skin far from the site of perforation of the parietal pleura (Fig. 21). By this the tissue duct can be compressed during the extraction maneuver of the drainage tube, until an accordingly tight sealing adhesive plaster (Opsite® oder Tegaderm® foil) is put on. In contrast to the previously used ointment swabs, these foils securely seal the drainage orifice and on the other hand the wound may be easily controlled because of their transparency at any time. Also during and immediately after extraction of the drainage tubes we asked the patients to hold some seconds in the exspirium, until the sealing foil is securely fixed.

12.1.1.19 Pseudarthrosis

See floating sternum.

12.1.1.20 Relapse/recidive

A relapse above all more often is to be expected with the conventional Ravitch method, than with the techniques using supportive metal or bioabsorbable struts or so-called pectus bars. In contrast to the suspension with a metal strut or bar lasting for 1–3 years, with the method of Ravitch the thoracic wall is tightened merely with absorbable materials, in analogy with a jumping sheet of the fire brigade. With decreasing strength of the vigor of the absorbable sutures or other absorbable osteosynthetic materials, the traction power of the diaphragm, the thoracic internal muscles and the negative pressure of the lungs regain their original strength. This strength is achieved before suitable biomechanical reshaping and enough stable cicatrizations of the ribs adjoining the remodelized area and the sternum have taken place. Recidivation in the techniques with pectus bar in children is rare [15, 20, 26, 29], if properly applied. However, relapses are to be expected when hardware for certain reasons (infection, allergy, other quarrelsomeness, and patient's wish) has to be removed prematurely and already after few weeks or months. Other reasons for a relapse are tilting of the pectus bar by too high-pressure load of the sternum [10] or insertion too laterally with transmuscular sloping along the intercostal spaces especially in adults or athletic adolescents. Adults with severe pectus deformities and asymmetric defects are at a greater risk of recurrence after a Nuss procedure. These patients may better be served with a modified Ravitch [19] or MOVARPE (Chapter 6.4) repair initially. Antonoff concluded that patients with failed open repairs would have better success with minimally invasive reoperations, whereas patients with failed minimally invasive repairs will have better outcome with open reoperations. When faced with reoperative pectus excavatum, these authors recommend consideration of an alternative operative approach from the initial procedure [2].
Late relapse of the deformity may occur upon pectus bar removal, if its bending was not yet slightly overcorrected at the time of implantation.

12.1.1.21 Rib non-union

Many ribs are cut or partially resected during several forms of thoracic remodeling procedures. Reefing sutures commonly approximate the cartilaginous rib stumps to such an extent that scarring forms a stable fibrous plate, occasionally with regrowth of cartilage tissue within. In rare cases, non-union, they may be caused by surgical overresection or by improper handling of immobilization braces through the patient, may result. Such non-union may lead to crepitation sensation, palpability, and audible cracking upon special movement, or even pain. Only in sequelae with permanent pain a reintervention seems to be justified then.

12.1.1.22 Scars

See Chapter 12.2.

12.1.1.23 Silicone implants-related problems

During fabrication of the mould, attention must be paid on the appropriate size and particularly in asymmetric cases on the orientation of the final implant. It is therefore recommended to document the site and orientation of the mould by photographs (Fig. 9b in Chapter 6.5).
Heavy weight implants by time tend to dislocate to caudal (Fig. 28) due to gravity and lack of suspension tissue within the subcutaneous plane. If such large blocks are required due to refusal to alternative methods, the manufacturing of implants with straps is recommended. Such straps are then fixed to the underlying skeletal structures with non-absorbable sutures to prevent further dislocation (Fig. 29).

Fig. 28. A horizontally placed silicone block to augment a caudally situated pectus excavatum deformity. Gravitation leads to displacement and unsightly deformity, due to lacking tissue suspension. The patient desired its removal

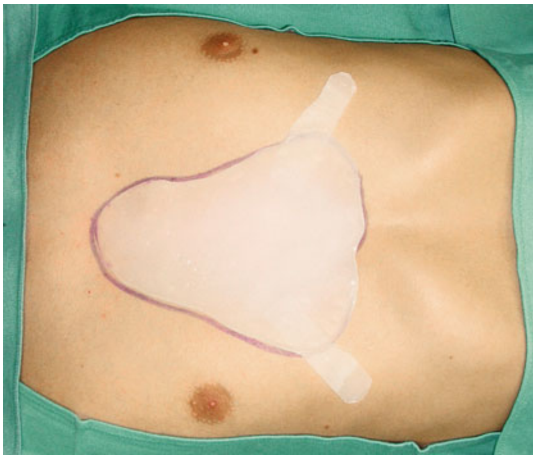

Fig. 29. Texturized and reinforced straps fixed to a custom-made silicon implant for fixation to skeletal structures to prevent dislocation by gravity or rotation

For typically silicone related but late problems as capsular contraction and others see Chapters 6.5 and 12.3. No history of cancer development from solid silicone parts having been implanted for decades is known. On the other hand, with silicone-filled breast implants single case reports exist which report the development of desmoid tumors associated with breast implants [3, 7, 18]. However also herein no evident relation between the silicone shell or the fluid components of the implant and the development of desmoid tumor can be derived so far, thus seems to remain a coincidental occurrence.

12.1.1.24 Tilting of the pectus bar

When a very rigid thoracic wall depression, as it is observed in young athletic males or adults, is elevated by one or even two pectus bars and when these pectus bars cannot not be fixed in the area of the sternum while using the MIRPE, the permanent pressure of the sterno-costal unit combined with perpetual breath excursions and physical movements successively force the pectus bars to dislocate from the horizontal plane. Besides, this tilting still after weeks and months takes place almost always without exception to cranial (Fig. 24a and b), because the intrathoracal insertion of the diaphragm to the sternum prevents dislocation to caudal. Such a tilting of the pectus bar can also be initiated and aggravated by lacking compliance of the patient, namely when the patient burdens the thorax and shoulder belts too early (Fig. 30a and b) exemplarily by body-building or other sportive activities. However, lacking technical comprehension when the pectus bar is bent in a too narrow radius and the over-elevated vertex

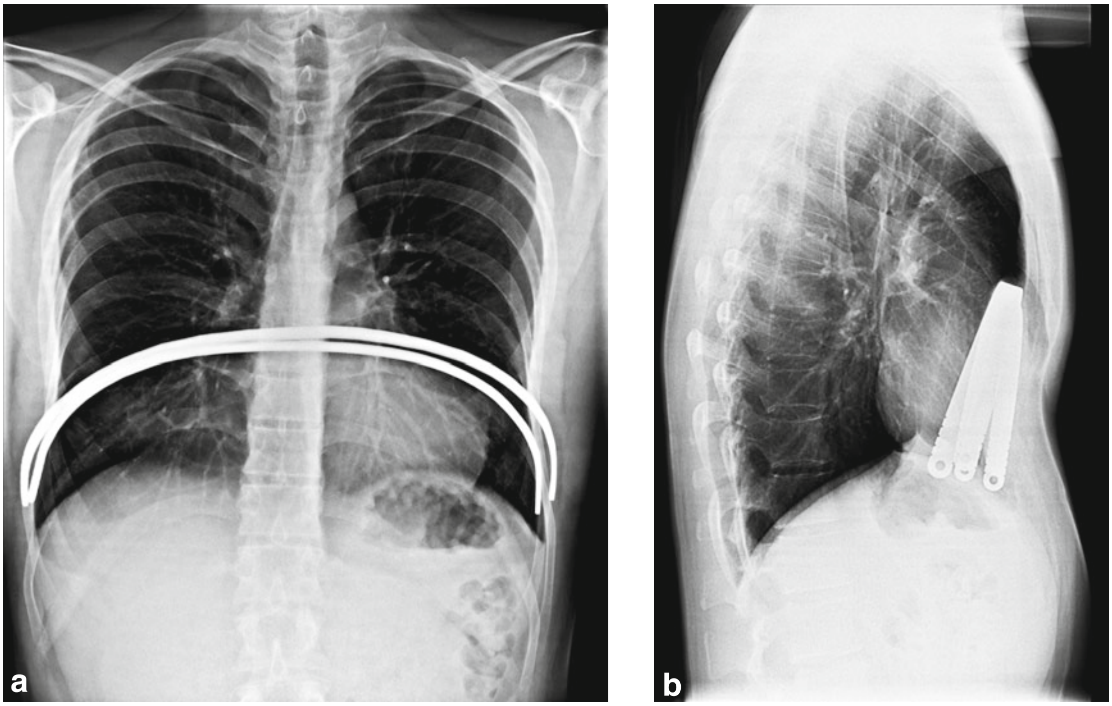

Fig. 30a and b. Completely tilted pectus bars few weeks after implantation. Despite instruction to rest the 34-year-old male went to perform body building immediately after demission from the hospital

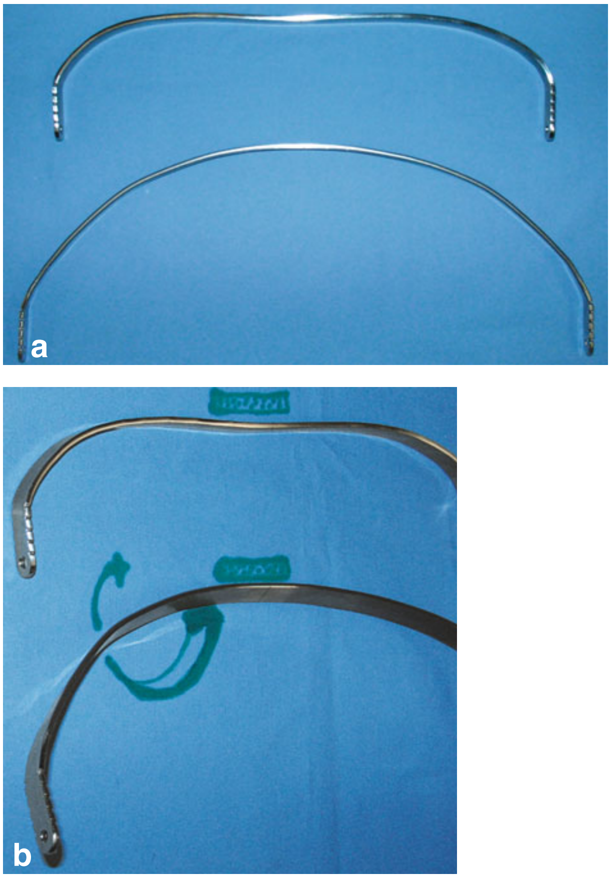

Fig. 31a and b. An "M" shape may prevent undue localized pressure at point to the pectus bar, minimizing the risk for tilting. On the other hand, sufficient bending forward must be present to enable slight overcorrection of sternal elevation to prevent long-term relapse. The lower pectus bar in **a** is projecting too much, endangering the stability of support, whereas the upper pectus bar seems to distribute the skeletal load much better along a flat course, but is bent too flat, hardly containing sufficient capacity to elevate a sunken sternum to a desired extent

(Fig. 31a and b) of the pectus bar comes to support the sternum merely in points can also cause a dislocation (Chapter 6.3). In all probability, this selective small spot of support, additionally if the pectus bar is not fixed there with sutures, by perpetual respiratory movements forces the bar to incline finally resulting in complete luxation. A possibility to convert this support in points to a flat area of support consists in a modified, slightly wave-shaped bending of the pectus bar (Fig. 31a). For these reasons some authors and we have abandoned the use of the lateral stabilizers in our institution, because they can only insufficiently compensate for the often immense pressure and lever force on the pectus bar. Instead, in the cases where strong pressure is to be expected (athletic disposition or adult), we prefer the combined semi-open access (MOVARPE) for the pectus bar implantation with sternum osteotomy and several circumcostal sutures [17] of the pectus bar fixed to the ribs in a three-point manner (Chapter 6.4). If in theses cases with circumcostal fixation, however very rarely a tilting still occurs, this may be due to surgical mistake that a fixation suture does not include the underlying rib [5]. During the MIRPE and MOVARPE techniques one should therefore inspect such circumcostal suturing by endoscopy in order to be sure that the sutures are well placed sparing and circumventing either the subcostal neurovascular bundle or the proper course around the rib itself (Fig. 27a and b).

12.1.1.25 Twisting (malrotation) of sternum

In an asymmetrical funnel chest deformity with twisting of the sternum out of the frontal (coronal) plane, the MIRPE solely, without adjuvant surgical maneuvers and rather probably in adolescents and adults inevitably leads to a new asymmetrical form of the thoracic wall, an iatrogenic pectus arcuatum deformity (Figs. 1 and 2a). Along with the pectus bar support of a twisted (malrotation) sternum in an athletic or elderly patient with inelastic cartilages, as well in preexisting arcuate deformity, this may create or even augment a pectus arcuatum appearance. A sternum malrotation in the child on the other hand can still be transformed relatively well to a flattened appearance by a pectus bar, conditioned by the elasticity of the sternum at the border between manubrium and corpus as well as the still pliable rib cartilages. Nevertheless, this is mostly entirely impossible in the adult, the older or athletic the patient is. Malrotations in these cases can be corrected only by an additional horizontal sternum osteotomy without or with parasternal rib cartilages resection or weakening (Chapter 6.4), depending on the extent and shape of the deformity as well as other influencing factors such as age, body size, and shape (athletic or leptosome). However, an excessive asymmetry, herein a deep unilateral depression can hardly be elevated to a symmetric shape, with all available methods and techniques or supportive devices, because of lacking support of the pectus bar on the affected side (Fig. 32). In many of these cases also a general hypoplasia of the affected hemithorax exists. Thus only a moderate improvement of the deformity can be expected, indeed, with more substantial invasiveness and increasing likelihood of perioperative complications or undesirable long-time effects such as relapse or unusual deformities in the area of surgical dissection.

Fig. 32. Severe asymmetric deformity with major volume deficit at the right hemithorax. Pectus bar implantation alone will result in minor improvement only. An invasive and complete remodeling, preferentially in several steps would be an alternative

12.1.1.26 Wound healing delay

Skin damage and soft part damage through inadvertent manipulation can delay the healing considerably from days until weeks. With ongoing healing delay, a surgical revision may be required in order to avoid exposure of any kind of alloplastic or metal implants. If ensuing aesthetically or functionally disturbing scars emerge, a correction is usually required. Simple scar correction in the pectus excavatum cases with strut suspension can be postponed and scheduled with implant removal. Very rarely, but especially with the use of silicone implants, stubbornly wetting fistula can emerge, which often heal up not before the implant is removed.

A 20-year survey with 1949 interventions follow-up was published in 2008, with collections of mostly children emerging out from 19 publications [33]. The results showed that most complications are related to the pectus bar itself in terms of dislocation, shifting, or others (37%). The second concern is pneumothorax (23%) or other pleuropulmonary problems (13%) followed by wound infections (14%) and pericardial effusion, and finally by hemato-thorax (4%). This study found no lethal incidents reported, however it is focused on a patient's collective with a mean age of 10.6 years. Many rare complications, most of them probably of minor nature may not be listed here, as they do not gain sufficient importance to be published in the medical literature, thus remain unknown so far. The list mentioned above may be a valuable information about experienced hence published complications but is unable to exclude other potentially more rare and so far unknown incidents.

An informed consent must include these most frequent complications as well as typical complications related to each individual type of surgical repair.

12.1.2 Peri- and postoperative complications in the keel chest surgery

Usually this intervention is technically much simpler than the invasive approaches of a funnel chest repair. Therefore complications occur to a much lower extent. The most frequent one may be laceration of the parietal pleura with ensuing pneumothorax or lesion of the internal thoracic artery and vein during parasternal cartilage resection, during sternum osteotomy, or during setting of the reefing sutures. After completion of the remodeling act and prior to final wound closure, the anesthetist is asked to raise the ventilation pressure (PEEP) up to 30 mmHg for a minute, while the surgical cavity is filled with saline. An eventual leakage of air will show air bubbles emerging from the site of pleura laceration. If the lesion is minimal, raised PEEP is maintained until definite and watertight wound closure. A small sickle-shaped pneumothorax, verified by X-ray immediately postoperatively may be neglected and usually will undergo spontaneous absorption within a few days. If present to a larger extent or showing respiratory impairment, a chest tube has to be inserted.

A postoperative chest radiogram is mandatory in every case anyhow; it may also detect intrathoracal hemorrhage or atelectasis due to pleural effusion timely.

Especially in extensive muscle flap elevation during the conventional Ravitch procedure and also in the modification with the muscle split technique, if muscle splitting is extensively performed to dorsal, damage to the pectoral branches of the brachial plexus might result in segmental muscle palsy with unsightly atrophic areas [37]. Such atrophies particularly in males may become a cumbersome unaesthetic flaw.

Wide undermining of skin flaps during surgery has to be avoided. Such undermining of skin flaps on the other hand is necessary especially when the muscle split technique is used to expose the affected ribs. In extensive keel chest deformities with required lateral exposure of the ribs therefore the conventional muscle flap elevation is preferred, since vascularization to the thoracic skin is still maintained through muscle perforator vessels. Wide undermining of such epimuscular skin flaps may cause perfusion problems that may be aggravated by the postoperative compression garments or keel chest brace paddle thus causing skin necrosis.

During the postoperative course, particular attention also has to be paid to the skin at the back around the implantation site of a peridural catheter. Infections there may cause migration of bacteria along the analgesics tube from the skin into the peridural space, eventually causing meningitis. As long as the peridural catheter is placed, the peripheral nerves and functions of the legs must be controlled, as the peridural catheter, however extremely rarely, may cause peridural bleeding with ensuing compression of the spinal cord, resulting in numbness or even paresis. Such complications need immediate consultation of a neurosurgeon and CT scans to be followed by instant treatment.

Hematoma at the site of remodeling usually does not occur because an elastic compression (cingulum) is applied and changed until it will be replaced by a custom-made keel chest brace after several days.

Single shot administration of broadband antibiotics prior to skin incision usually is sufficient to avoid however uncommon wound infections.

From the day a postoperative keel chest brace is applied, the skin underneath the compression paddle at the site of remodeling as well as underneath its counterpart on the back has to be inspected daily in order to detect and circumvent major pressure sores or skin blisters (Fig. 33) which might result in skin necrosis and unsightly scars (Fig. 34).

In order to avoid recidivation or demodeling of the anterior thoracic wall, the patient must be instructed

Fig. 33. Superficial pressure sores from too tight application of a postoperative keel chest brace

Fig. 34. Undue pressure on a keel chest brace led to superficial skin necrosis and as a consequence to a well visible scar

thoroughly about how to handle this device during the period of 6–8 weeks, It is therefore recommended to supervise the patient for 1 or 2 days prior to demission and call him/her back as out-patient within several days to control the appropriate handling.

Late complications may occur after inappropriate or inconsistent application of the keel chest brace. This may result in minor (Fig. 35) or major recidivation of the keel deformity due to lacking support of the healing process by pressure. The brace furthermore supports the

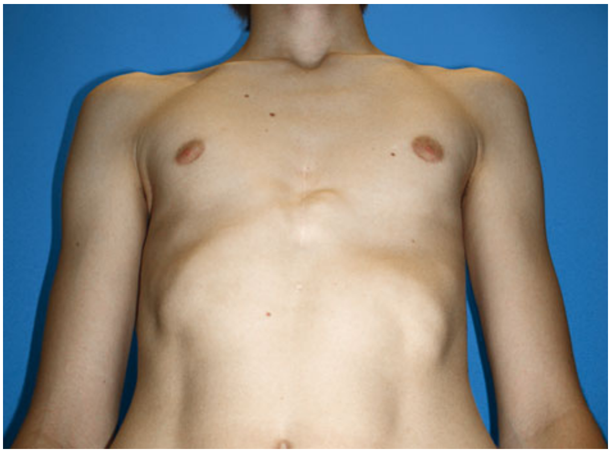

Fig. 35. Minor regrowth of rib cartilages 1 year after surgery of extensive keel chest deformity

healing of the skin incision with regard to scar compression and avoidance of hypertrophic scar formation (Chapters 7.4 and 12.2).

12.1.2.1 Postoperative treatment

Physical condition training without stressing the shoulder girdle should be possible immediately after removal of skin sutures; increased physical load with muscle training may be recommended in children after 4–6 weeks, in adolescents and adults not prior to 6–8 weeks. Maximal physical load with excessive sports activities should be postponed for 12 weeks to allow secure healing of implanted metals or other alloplastic material. After the time of immobilization, an internal scar with a strong layer of fibrosis will definitely prevent the pectus bar from any dislocation [5].
Within minor surgical corrections these periods may be shortened according to the technique applied and the invasiveness of surgery, as well as to age and the patient's intellect and compliance (see relevant chapters).

12.1.3 Conclusion

To the prerequisites in the treatment of such a variety of interventions belong experiences with the treatment itself and furthermore with the follow-up treatment, well-instructed nursing personnel as well as physiotherapists. The availability of an intensive care unit, interdisciplinary networking, and co-working in the emergency case is mandatory prior to undergoing invasive correction surgery at the thoracic wall. Only with familiarity in the utilization of different treatment methods, a high operation frequency and corresponding routine of the surgeon, the intervention remains relatively without risk and can lead to high success and content patients.
Complications overall are dependent on many factors, such as the technique used, the patient's age, and the experience of the treating surgeon. Generally only in exceptions, e.g. in severe deformities and demonstrable functional impairment of the intrathoracic organs, interventions may be justified already at an early age. Otherwise surgery should be performed better within or after puberty due to better understanding with the patient and an already finalized shaped deformity by growth maturation [16].
Taking into account not only the frequent [16, 26, 29, 33] but also the typical and specific complications, a complete and thorough informed consent must contain such information. The thoracic wall deformities in approximately 90% ostensibly represent aesthetic problems, however with more or less psychic components. Therefore a much more detailed enlightenment with respect to results that reach from undesired outcome with aesthetic flaw only until life-threatening incidents is mandatory during the conversation with the patient who seeks treatment. The publication of adverse effects or complications is not a rewarding task and therefore such reports usually are available to a limited extent, but they are immanent during every such often highly sophisticated procedure. Particular and furthermore detailed knowledge as well as keen awareness of these potential complications, even of the rare ones is required in order to keep the learning-curve flat and to avoid personal experiences with injured patients and the court.

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12.2 Hypertrophic scars and keloids

Dolores Wolfram-Raunicher

12.2.1 Background/introduction

Hypertrophic scars (Fig. 1) and keloids result from an abnormal wound healing process in which control over tissue repair and regeneration is lost. Such an abnormal fibrous growth presents a major therapeutic challenge and sometimes dilemma to the physician because these particular scars are disfiguring and frequently recur. In general, patients present with aesthetic concerns, however hypertrophic scars and moreover keloids can also cause pruritus, pain or pressure [1].

Fig. 1. Adolescent with hypertrophic scars 1 year after pectus bar removal and additional scar correction at the xiphoid region. The hypertrophic scar remains despite silicone sheet compression over 2 months. The xiphoid scar resulted from mini-open thoracotomy for the digital guidance of pectus bar insertion 4 years ago, utilizing a modified MIRPE technique

12.2.2 Differential diagnosis of hypertrophic scar versus keloid

Hypertrophic scars and keloids are defined as abnormal wound responses in predisposed individuals and display an overregulated connective tissue response to trauma, inflammation, surgery, or burns [2]. The first step to optimal scar therapy starts with the simple identification and diagnosis of the aberrant scarring [3]. A "hypertrophic scar" as defined by Peacock in 1970 [4], is a scar elevated above the skin surface level that stays within the margins of the original wound. The term "keloid" is used for a scar, which also is elevated above the skin surface level, but proliferates beyond the margins of the original lesion. Hypertrophic scars usually subside with time and are painless, whereas keloids continue to evolve over time, without a quiescent or regressive phase [2–5] and can present with itching and pain.

12.2.3 Etiology

There are many possible factors resulting in abnormal wound healing. Genetic predisposition, skin or wound tension, infections and incisions beyond the relaxed skin tension lines are the most important ones [2, 5]. Scars crossing joints or skin creases at a right angle are predisposed to form hypertrophic scars due to the constant tension forces that occur [6]. Moreover, specific anatomic sites, such as the sternal region, the shoulder and the upper part of the back are typical regions to produce abnormal scarring. These regions are characterized by higher skin and muscle tension, and the lack of subcutaneous tissue especially in the sternal region influences the abnormal wound healing in a negative way. Previous studies in literature have shown that median sternotomy hypertrophic or keloid scarring is prevalent among cardiac surgery patients. The incidence of abnormal scarring in this region is approximately 30% in the Caucasian population [7]. Keloids can occur at any age, however, they often develop during and after puberty [2]. Davies explained this prevalence by the fact, that younger individuals are more frequently subjected to trauma and their skin is more tight than the skin of elderly persons [8]. The observation that keloids are 15 times as likely to occur in darker skinned individuals points to a genetical background [9]. Keloid formation mainly occurs in parts of the body with high concentrations of melanocytes, whereas it is rare on the sole and palm. Keloid formation has also been associated with endocrine factors and menopause seems to prompt the recession of keloids, whereas pregnant women report keloid onset or enlargement [10].

12.2.3.1 Pathophysiology of hypertrophic scars and keloids

The development of a normal scar involves the influx of inflammatory cells into the wound, and the production of growth factors for the wound healing process (e.g. transforming growth factor-beta; TGF-β) [5]. In response to these factors, fibroblasts migrate into the wound and synthesize collagen. During normal wound healing, the nodularity and redness of the wound soften and flatten due to ongoing simultaneous collagen synthesis and degradation; the connective tissue elements regress after the third week [2, 5]. This balance of matrix degradation and collagen biosynthesis is disturbed in keloids and hypertrophic scars, resulting in excessive accumulation of collagen in the wound. Literature shows, that collagen synthesis in keloidal tissue is approximately 20 times greater than in normal unscarred skin and three times greater than in hypertrophic scars [11, 12]. Aside from high collagen synthesis and proliferation of fibroblasts in keloids, other authors found that keloid-derived fibroblasts show a rate of fibronectin biosynthesis that is four times higher as that of fibroblasts from normal scars and normal dermis [13, 14]. Less synthesis of molecules that promote matrix breakdown may also explain the lack of scar regression seen in keloids [15].

12.2.3.2 Treatment for hypertrophic scars and keloids

The treatment of hypertrophic scars and keloids can be subdivided into surgical, nonsurgical, and a combined modality approach. Unfortunately, up till now there exists no therapeutic standard concept for abnormal scarring due to the heterogenous etiology. Moreover, only few studies provide a coherent therapeutic plan because of poorly defined endpoints in treatment, lack of prospective studies, inadequate follow-up care, and failure to properly distinguish between hypertrophic scars and keloids. However, most of the literature on keloids suggests high recurrence rates, regardless of the modality of treatment [16].

Surgical treatments

(a) Surgery for keloids

In keloids, simple total excision stimulates additional collagen synthesis, hence often prompting quick recurrence of a keloid probably even larger than the initial one [16, 17]. Therefore, intralesional surgical excision of keloid tissue is recommended [18]. Since surgical excision of a keloid is associated with a high recurrence rate [19, 20], surgery should be combined with adjuvant treatment such as pressure or intralesional corticosteroid injections. Nevertheless, the surgical therapy for the treatment of keloids has been relegated mainly to a second-line therapy for lesions unresponsive to steroids or pressure [11].

(b) Surgery for hypertrophic scars

Excision is the main therapy for patients with hypertrophic scars from complicated (e.g. infected) wounds or delayed closure. Two aims are achieved by scar revision: excision and narrowing of scars likewise to widespread scars and the appliance of a Z- or W-plasty to alter the direction of the scar [2], so that the new scar lies within the relaxed skin tension lines [21].

Nonsurgical treatment modalities

(a) Topical silicone gel

Application of topical silicone gel sheets is very popular in the treatment of abnormal scarring, although the mechanism of action is unknown. It has been suggested, that increased wound hydration affects local keratinocytes to alter growth factor secretion and to influence fibroblast regulation [22, 23]. It is also assumed that increased hydration decreases capillary permeability, inflammatory and mitogenic mediators, and reduces collagen synthesis [24]. In patients prone to hypertrophic scar formation, topical silicone gel sheeting has a distinct effect in impeding the formation of abnormal scarring after surgery. Application of silicone gel sheets should start as soon as reepithelialization of the wound is finished, and daily application for at least 12 h is recommended [25].

(b) Corticosteroids

Intralesional corticosteroid injections represent a main therapy in the treatment of abnormal scarring, alone or in combination with other therapeutic procedures [26]. The intralesional application of corticosteroids can soften and flatten keloids but cannot narrow hypertrophic scars or eliminate keloids [2]. Corticosteroid injection leads to decreased fibroblast proliferation, collagen synthesis, and glycosaminoglycan synthesis and also suppresses pro-inflammatory mediators [24, 27]. Steroid injections must be administered cautiously to avoid overtreatment, which may result in skin and subcutaneous fat atrophy, teleangiectasy, and a depressed scar. Additionally, the use of corticosteroids in children is controversially discussed due to possible systemic side effects (e.g. growth suppression, hyperglycemia, and Cushing syndrome) and the painful injection procedure.

(c) Laser therapy

Laser therapy has been tested for the treatment of hypertrophic scars and keloids with varied results [28, 29]. Superiority of laser use over simple excision of keloids currently has not been demonstrated in clinical trials. However, some authors reported promising results, using the pulsed dye laser (PDL) for the treatment of hypertrophic scars. Alster reported an average improvement of 57% after the first and 83% after the second treatment with PDL for hypertrophic and traumatic scars [28, 30]. In addition to the reduction in erythema, flattening, a reduction in itching and pain, and optimization of the skin texture have been observed in hypertrophic scars [30].

(d) Pressure therapy

The use of pressure to treat keloids was already described in 1835 [24]. However, pressure therapy was not popularized until the 1970s, when physicians noted that pressure stockings used on lower extremity burns resulted in less erythematous and thick scars [31]. The compression phenomenon is not well understood, but the application of pressure leads to a decreased blood flow in the treated area, which favors collagen break down and thus softens keloids. Additionally the pressure-induced hypoxia involves

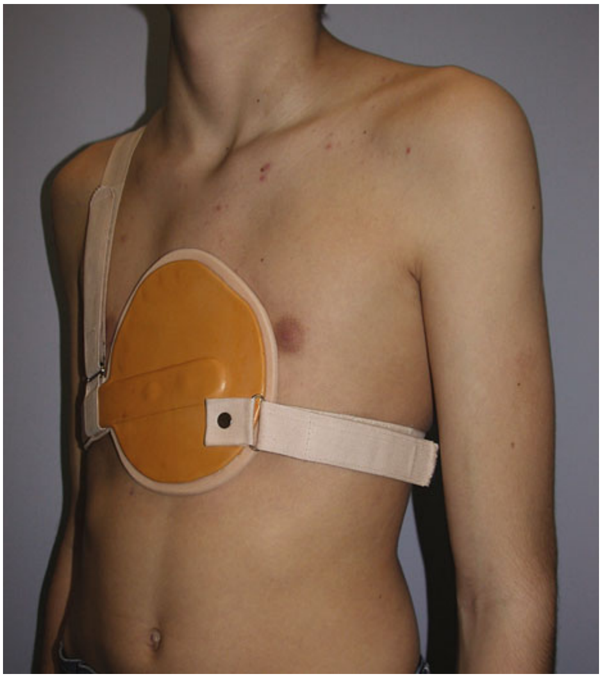

Fig. 2. A keel chest-brace with a silicone foam pad, armed with an elastic metal strut which is covered with artificial leather. The pad covers the whole area of cartilage and sternal bone remodeling, additionally performing intrinsic compression on the scar

Fig. 3. Fourteen years old boy with Marfan-syndrome. Nine month prior to keel chest remodeling aortic surgery was performed with a resulting hypertrophic scar all along the midsternal incision. After the keel chest surgery and excision of the whole scar, compression was applied only above the area of skeletal remodeling. Fourteen months thereafter the compressed scar clearly visibly shows minor hypertrophy in contrast to the non-compressed part cranially and to a small part at the epigastrium

fibroblast degeneration. Pressure therapy should be started immediately after reepithelialization of the wound, and therapy must be instituted for long periods (>23 h/day for up to 6 months) before significant effects may be achieved. Unfortunately, many regions of the head and neck are not amenable to pressure application. Additionally, the success rate depends largely on patient compliance. Fortunately and particularly in the keel chest deformity, orthopedic technicians provide with a so-called keel chest brace (Fig. 2). That device is applied immediately within days after remodeling surgery and in almost all cases of our personal experience (Chapter 7.4) prevented hypertrophic scarring or keloid formation [32]. The hypothesis underlying that beneficial effect is, that pressure applied already during wound healing and at the very beginning of scar formation, for 2 months, diminishes collagen synthesis through reduced vascularization (Fig. 3).

Conclusion

The development of hypertrophic scars and keloids still represents a frustrating problem for the patient and the physician as well. Patients and their parents need to be informed about the risk of abnormal scarring, especially in the sternal region. Additionally, we recommend closely monitored postoperative controls at an experienced specialist to early diagnose an abnormal wound healing and to initiate optimal treatment in an adequate early phase. In keel chest patients a postoperative keel chest brace not only maintains the reshaped thoracic wall but also prevents unsightly scar formation. In the other collection of thoracic wall deformities, the routine consideration of incisions along the relaxation skin tension lines and beyond the decolletée is of prophylactic importance.

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12.3 Silicone implants and their features

Dolores Wolfram, Evelyn Rabensteiner

Breast augmentation with silicone mammary implants is still one of the most commonly performed procedures in plastic surgery [1]. Despite the controversy about the safety and efficacy of silicone gel implants [2, 3], women's desire for perfect formed tight breasts has led to a boom in aesthetic breast augmentation. The most common indications for breast augmentation are hypoplasia or aplasia of the breast, breast reconstruction due to cancer or fibrocystic mastopathy, and congenital breast deformities like tubular breast and asymmetry [4]. Moreover, silicone implants are also used for the aesthetic correction of thorax deformities, namely funnel chest, keel chest or Poland syndrome.

Currently, the two implant types used are silicone gel filled breast implants or saline filled implants, which both feature a highly polymerized silicone shell. Both implant types may be smooth or microtextured at their surface. The common breast implants are either spherical or anatomical in shape [5]. However for the correction of deformities like funnel chest or asymmetrically depressed ribs, custom-made silicone implants are available [6]. In the latter case, the implant is individually designed based on the extent and shape of the thoracic deformity. These implants, in contrast to breast implants with a shell and saline or gel filling, consist of highly polymerized silicone throughout (Chapter 6.5).

Although serious complications in context with silicone implants are rare, potential problems can be divided into early and late complications. The early complications are due to the surgical procedure (e.g., hematoma, seroma, infection, asymmetry), and the later are based on the body's response to the implant itself. The late complications include capsular contracture, deflation or rupture as well as systemic complications [2, 7, 8]. A fibrous tissue reaction develops around every type of implant or foreign body, however the term capsular contracture includes formation of a hypertrophic fibrotic capsule around the implant that is prone to contraction in later stages, causing severe pain, local tissue damage and implant function impairment [9, 10]. Capsular contracture remains the most common complication associated with silicone breast implants, occurring in 4–11% of women [8, 11]. However, literature is very inconsistent regarding exact incidence rates due to variable observation periods, different implant types and inadequate follow-up [8, 11]. Aside capsular contracture, implant rupture and silicone leakage are reported in literature [12–15]. Silicone leakage can not only lead to formation of siliconomas within the capsular tissue or breast, but also induce lymphadenitis and inflammatory responses in other organs [13, 14]. Implant rupture rates in literature are varying between 1%

Fig. 1. Chronic inflammation of a two-component silastic rubber device, implanted 22 years ago. The sterile inflammation, confirmed by aspiration and microbiological examination, led to massive seroma formation and pain

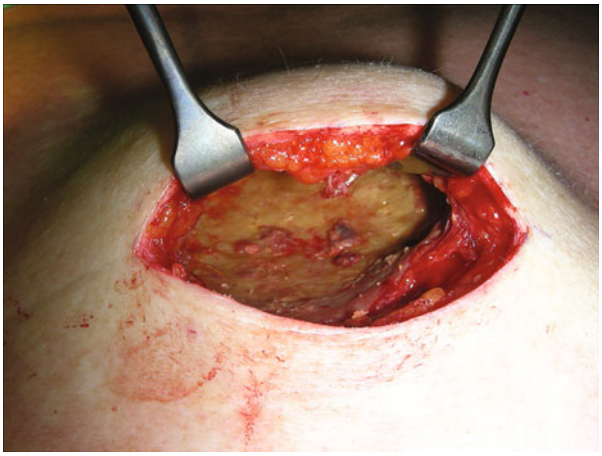

Fig. 2. Same patient as in Fig. 1. After extraction of the silicone rubber material the inner surface of the capsule revealed a calcificated shell with abraded silastic mucus

and 17% depending on the retention time of the implant within the body [15].

In addition to these phenomena, several case reports of systemic connective tissue diseases [16–20], primarily scleroderma-like or rheumatologic symptoms and musculoskeletal disorders, mainly in women with ruptured breast implants have been published. Meta-analyses were performed worldwide to clarify the controversial relation between silicone mammary implants and systemic side effects [21–23] but the majority of these studies have neither confirmed or denied an absolute safety of silicone breast implants.

Although silicone breast implants are considered as safe medical devices and some manufacturers even postulate lifetime guaranty of their silicone breast implants, patients need to be informed about early and late side effects and thus the need for an implant exchange or removal, irrespectively if filled with saline, with gel, or solid in structure (Figs. 1 and 2).

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Actual technical improvements, future aspects

13

13.1 Thoracic wall deformities: 3-D scanning and computerized remodeling

Laszlo Kovacs, Maximilian Eder, Christian Brossmann, Anton H. Schwabegger

13.1.1 Background

The morphological estimation of complex anatomic body regions and of congenital or acquired defects is of great importance for an individualized surgical planning, the operative quality management and for long-term postoperative outcome control [11]. Especially congenital thoracic wall deformities frequently show substantial anterior thoracic wall affections, but only few and often deficient analyzing methods exist to objectively assess the complex three-dimensional (3-D) contour anomalies [19]. In particular, women show associated malformations of the breast soft tissue, which aggravates the 3-D evaluation of the female breast region.

In the last decades different evaluation methods have been presented to classify the severity of an anatomical distortion to enable a more objective quantification of an anterior chest wall depression [3]. Direct linear body surface measurements or radiologic distance measurements (chest X-ray, computed tomography (CT), nuclear magnetic resonance imaging (MRI)) provide specific anthropometric indices to quantify chest wall deformities and to compare the surgically induced contour changes [1, 5, 9, 10]. The generated indices only measure the present body deformity punctiform and along one body axis, they are difficult to standardize and show examiner-dependant measurement inaccuracies. Sometimes even invasive examinations are needed and thus inadequately assess the defect geometry [11].

Conventional documentation and assessment of body surface changes using two-dimensional (2-D) photography also neglect spatial dimensions of the chest wall deformity and because of perspective distortion, lack of metric and 3-D information are not suitable and limited by empiric and subjective examiner interpretation [12]. Modern imaging procedures such as CT and MRI enable reconstruction of the complete thorax with all anatomical structures and also the body surface in three dimensions to provide a more vivid visualization and interpretation of the entire complexity of thoracic wall deformities. Highly disadvantageous herein is the patient acquisition in supine or prone position with resulting soft tissue deformation of the whole breast region, time and cost intensive acquisition and the invasive character of these techniques which disallows a routine application for clinical follow-up examinations at close intervals [8, 18].

This lack of criteria oriented clinical assessment methods raises the necessity for supplement existing techniques with additional evaluation methods to guarantee fast, non-invasive, routine clinical follow-up examinations at close intervals and objective, patient specific 3-D assessment and quantification of the body geometry.

New optical measurement systems for the 3-D body surface assessment fulfill the above named relevant requirements and different clinical applications in the field of plastic, reconstructive and aesthetic breast surgery have been demonstrated [16, 17]. One particular advantage of 3-D surface imaging is the non-invasive patient acquisition which provides a risk-free, non-contact, non-deformable, high-resolution 3-D virtual colored model creation of different and specific body regions. 3-D surface imaging enable precise and accurate pre- and especially postoperative 3-D quantification of shape, volume, surface changes, symmetry, projection, swelling tendency, contour and deformation, close intervals postoperative follow-up examinations and therefore optimizes specific steps in the preoperative surgical planning [7]. Digitalized 3-D datasets allow a well-structured patient documentation and computer-aided analysis and evaluation without the necessity of the patients being physically present. The possibility to work with the stored 3-D data in absence of the patients avoids time consuming patient measurements and enables the surgeon to work with different data versions for different clinical questions in different software solutions and to access, revise and overwork the data at any time, in particular even during surgery [14, 16–18].

Optical systems are based on the measurement principle of triangulation and consist of a transmitter unit (light source) which projects a point upon the measured object and a receiver unit (camera) which detects the reflected light. The geometric configuration and the measured angle between the transmitter and the receiver allow the calculation of the correct object point in space and the resulting surface geometry [2]. The measured surface is captured as an accumulation of different single points (point cloud). Every point is defined and computed on the basis of the respective x-, y-, and z-coordinates and provides mathematic precise visualization of the whole body surface [16, 17]. A wide range of optical systems for 3-D surface geometry acquisition exist and are based on different physical principles (stereo(-photo)grammetry, Moiré topographie, pattern/structured light, linear laser scanner, etc.). All optical surface imaging systems have in common that they detect the body surface in a short time without any deformable body contact to create a virtual digitalized 3-D model for later computer-aided analysis. In preliminary studies different body surface imaging systems were tested regarding their potential medical application and found the 3-D linear laser scanner most suitable for clinical applications [4].

By means of different patients with pectus excavatum deformities the vantages and existing limitations of current 3-D visualization techniques regarding objective 3-D quantification, computer-aided surgical planning and the development potential in the field of surgical correction of thoracic wall deformities are presented in the following.

13.1.2 3-D quantification of the body surface geometry

A common challenge in plastic and reconstructive surgery is the correction of congenital or acquired deformities. Precise detection and quantification would provide more security to surgeons during the intervention and would enable an objective quality assurance and thus would ease communication with the patient as well. The body surface geometry is quantifiable using 3-D surface imaging systems. The pre- and postoperative assessment of the breast and thorax region can be accomplished sufficiently precise and accurate using a 3-D linear laser scanner according to a standardized scanning protocol and can be presented as a virtual 3-D model for further computer-aided evaluation with appropriate software [16, 17].

On the one hand the accuracy of the existing scanning systems depends on the capturing speed; on the other hand accuracy depends on the capability to assess larger body regions in one or more steps. Current scanning systems vary enormously in fulfilling these requirements. To capture the whole anatomical region of interest it is often necessary to accomplish multiple surface scans from different angles to capture complex body regions. The capturing process for one single shot takes 1–2 s. More problematic is the creation of a closed 360° 3-D model. Especially congenital thoracic wall deformities are often associated with vertebral column deviation and a 360° scan is helpful to evaluate the deformation in total. To achieve a 360° view, multiple single shot scans must be performed either by changing the patient position according to the fixed scanner or by turning the scanner around the patient when using one single scanner. More complex scanning systems with multiple combined scanning modules are expansive but allow capturing a 360° view model with one single shot. Depending on the scanning system the whole capturing process takes from seconds until several minutes.

The 3-D quantification of the body region in a Cartesian coordinate system allows the creation of a virtual 3-D surface computer model using different algorithms and software. This virtual 3-D model offers several advantages over conventional 2-D photography: beside color information, spatial computation of individual points is possible on the basis of the respective x-, y-, and z-coordinates and virtual rotations of the 3-D model in all three dimensions. Precise and accurate pre- and postoperative volume, surface, distance and symmetry measurements on the virtual 3-D model in absence of the patient are feasible. Multiple visualization possibilities and clearly well-structured documentation of the 3-D shape differences allow a meticulous analysis of the anatomical region of interest. After interactive labeling of specific anatomic breast landmarks it is possible to automate some of the analyzing procedures and to release an automatically generated analysis report.

Several important software analysis functions for the clinical application are demonstrated in Fig. 1.

In contrast to conventional 2-D photography, linear measurements can be performed directly on the body surface using virtual 3-D models and clinical questions regarding breast projection, ptosis, circumference, etc. can be documented and visualized very precisely (Fig. 1a). Even surface anthropometric measurements described in the literature to provide severity indices of the deformity can be performed on a virtual 360° surface model (Fig. 1f). But these anthropometric surface measurements deliver only limited information regarding the complex 3-D deformity. The correct deformity degree regarding breast volume and breast surface deficits (Fig. 1b and c) can be quantified [8, 16, 17]. Further it is possible to compare the whole chest wall region

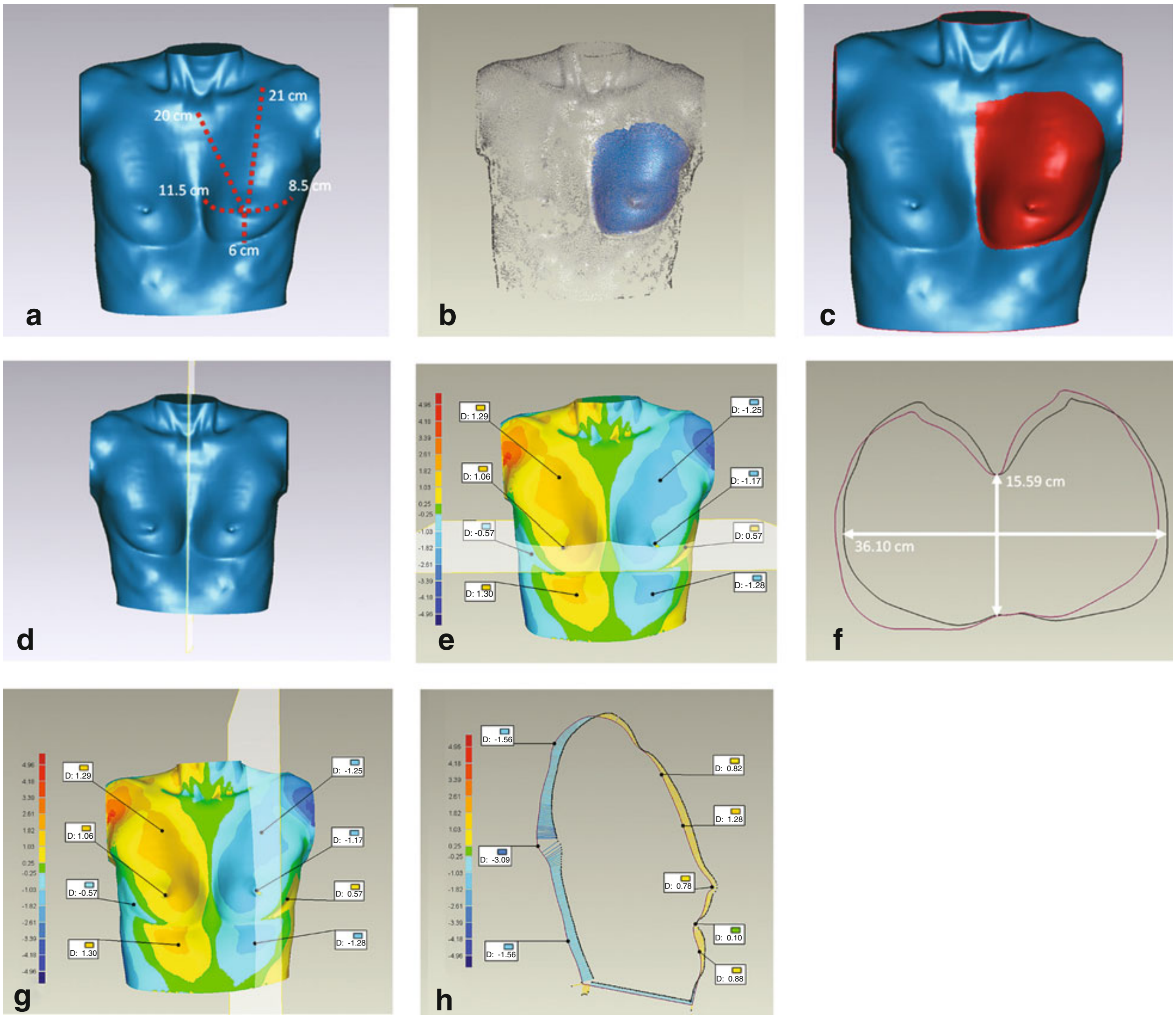

Fig. 1. Different 3-D quantification methods of the thoracic wall: **a** linear on surface measurements; **b** volume measurements; **c** surface measurements; **d** placing symmetric mirror plane; **e** and **g** color-coded quantification of thoracic wall asymmetry by superimposed mirrored 3-D model on the non-mirrored 3-D model and 2-D deviation slice placements; **f** horizontal slice with 2-D deviation of the two models and anthropomorphic thoracic wall measurement at nipple-areola-complex height; **h** sagittal slice between the left and the right thoracic wall with color-coded 2-D deviation at nipple-areola-complex height. All measurements in cm

surface by mirroring an anatomic region of interest (Fig. 1d) for an evaluation of the symmetry of the thoracic wall. By mirroring the 3-D model at the height of the sternum and by superimposing the mirrored model onto the non-mirrored model, precise color-coded quantification of existing or remaining asymmetries of the whole body surface are possible (Fig. 1e and g). Additionally, by placing different slices through the virtual 3-D model (Fig. 1f and h), asymmetries of the contra lateral breast region are visualizable [7, 14]. The visualization of 3-D and 2-D deviations, volume and surface differences etc. deliver important data for an improved preoperative planning of the surgically induced shape transformation [14]. The possibility to compare complex 3-D geometries also permits to superimpose pre- and postoperative 3-D models of a patient and to objectively quantify and evaluate the postoperative results [7]. 3-D surface imaging enables a routine follow-up examination at close intervals and is perfectly suitable for postoperative long time analysis regarding postoperative swelling and soft tissue shape changes over time by superimposing 3-D surface scans accomplished at different examination dates postoperatively. The summary of all these analyzing software functions in an automatically generated report allows quantification of the body surface to optimize surgical procedures as well as an objective quality assurance of the postoperative result and long-term postoperative follow-up studies.

13.1.3 Computer-aided surgical planning

Among others "computer-aided surgery" is aiming at supporting the surgeon during the preoperative planning process and even during the surgery. Through this perspective the influence of existing possibilities and limitations of 3-D imaging and preoperative analysis of the virtual 3-D models concerning the surgical planning process are demonstrated. Most patients undergoing

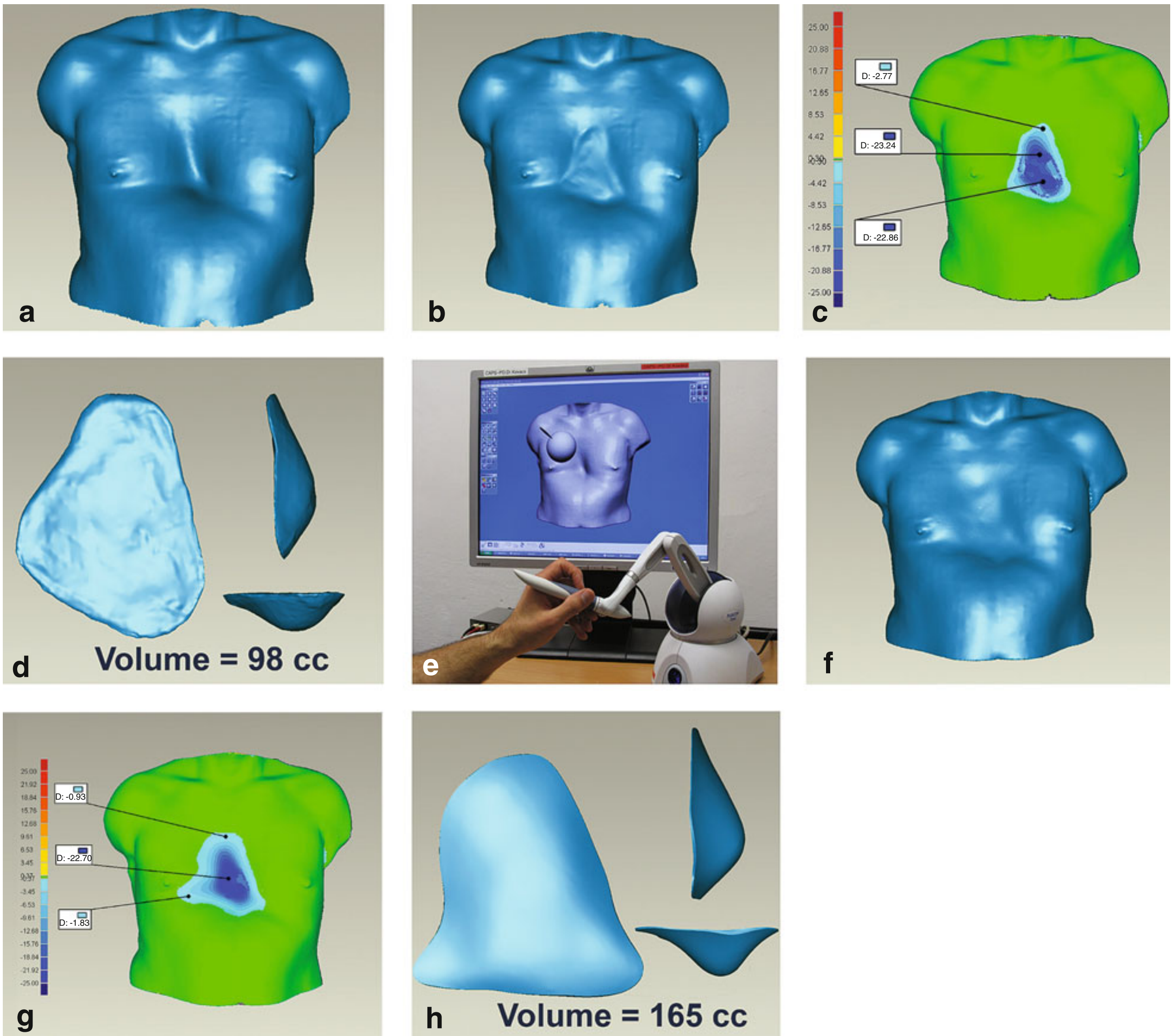

Fig. 2. Computer-aided planning of an silicone implant for chest wall deformity correction: **a** patients preoperative 3-D surface scan; **b** 3-D surface scan with attached conventionally planned silicone implant; **c** 3-D comparison of preoperative 3-D scan and 3-D scan with attached implant; **d** 3-D scan of the conventionally planned silicone implant; **e** force-feedback system for intuitive 3-D haptic interaction with the virtual 3-D model; **f** haptic deformed virtual 3-D model; **g** 3-D comparison of preoperative 3-D scan and haptic deformed 3-D model; **h** computer-aided custom-made silicone implant design by subtracting pre OP scan from deformed 3-D model. All measurements in mm

surgery strongly desire to anticipate the postoperative outcome prior to surgery. Specific software application systems which allow direct interaction of the examiner with the virtual 3-D model enable the surgeon an intuitive virtual 3-D modeling. In analogy to 2-D photography morphing systems it is possible to interactively modify the virtual 3-D model according to the patients and surgeons perception. By means of intuitive modeling of the virtual 3-D model the desired final shape can be displayed. In contrast to conventional 2-D photography not only a visual impression is presented but also concrete comparable measurements for the preoperative planning can be derived from such a 3-D intuitive modeling (Fig. 2).

The degree of thoracic wall deformities, especially of pectus excavatum, is extremely variable and according to their severity requests different reconstruction methods. Severe and symptomatic deformities are corrected with chest wall remodeling, removing affected costal cartilages with sternal repositioning and more mild and asymptomatic deformities can be reconstructed satisfactorily with subcutaneous custom-made silicone implants.

Using the example of a male patient (age: 37 years, height: 194 cm, weight: 100 kg, pectus severity index (PSI): 1.9) with asymptomatic pectus excavatum deformity (Fig. 3), the computer-aided surgical planning of silicone implant contour reconstruction is compared with the conventional planning method using the moulage technique (Fig. 2).

The implant is designed from a moulage taken of the chest wall deformity with the patient in the supine position. The moulage designed implant was 3-D assessed and a virtual 3-D model (volume = 98 cc) of the

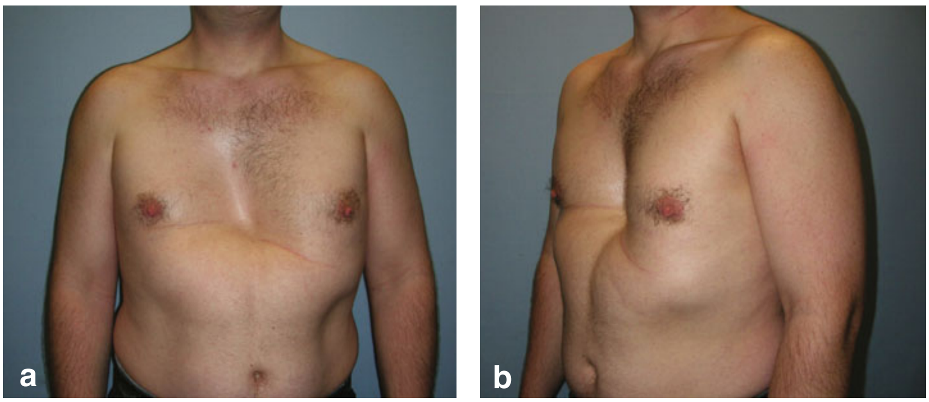

Fig. 3. Preoperative 2-D photography of an asymptomatic 37-year-old male patient undergoing pectus excavatum correction with custom silicone implant: **a** frontal and **b** oblique view of lower third sternal contour deformity

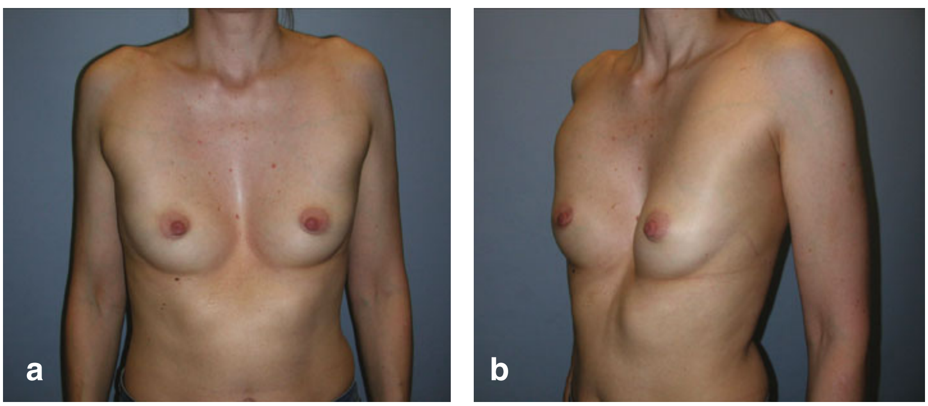

Fig. 4. Preoperative 2-D photography of a symptomatic 42-year-old female patient undergoing pectus excavatum correction with half open Nuss procedure and sternal osteotomy: **a** frontal and **b** oblique view of the anterior thoracic wall contour deformity

implant was created (Fig. 2d). Afterwards a preoperative 3-D surface scan of the patient's anterior chest wall was accomplished without and with attached moulage designed silicone implant (Fig. 2a and b). Both 3-D models were superimposed and a so-called 3-D compare with color-coded visualization of the existing differences of the two 3-D surface scans to each other was carried out (Fig. 2c). In a second step the preoperative 3-D surface

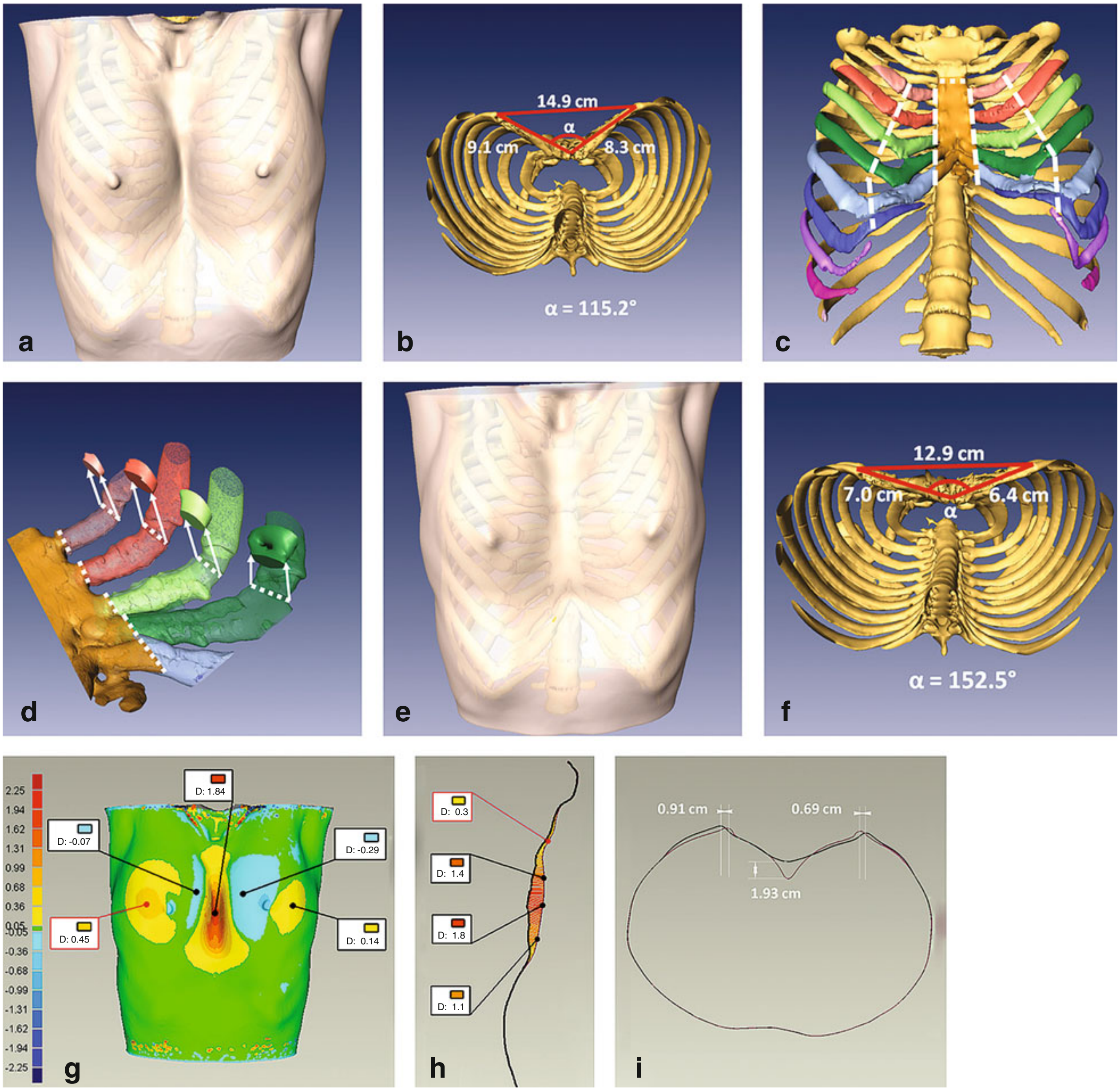

Fig. 5. Computer-aided planning of surgical thoracic wall deformity correction using 3-D CT scan data: **a** preoperative 3-D model of skin surface and deformed bony thoracic wall; **b** preoperative 3-D reconstructed bony thorax with linear and angle measurements of the deformation; **c** preoperative 3-D bony thorax with marked osteo- and chondrotomy excision lines (white dashed lines); **d** quantification of the excised osteotomy segments; **e** 3-D model of skin surface and 3-D bony thorax after virtual correction; **f** 3-D reconstructed bony thorax with linear and angle measurements after virtual correction; **g** 3-D comparison of the virtual corrected versus preoperative 3-D CT skin surface model; **h** quantification of thoracic wall shape change between pre OP (—) versus corrected 3-D CT skin model (—) using color-coded sagittal 2-D deviation slice at height of the sternum and **i** using horizontal 2-D deviation slice at height of the nipple-areola-complex. All measurements in cm (© CAPS/Brossmann)

scan (Fig. 2a) was interactively modeled using specific 3-D morphing systems (Fig. 2e). The postoperative soft tissue change for the reconstruction of the bony contour deformity after silicone implant correction is directly modeled intuitively on the virtual 3-D model by the surgeon and can be precisely quantified and visualized (Fig. 2f). The precise volume and the correct shape of a conventionally designed silicone implant can also be virtually designed (Fig. 2g). The virtually designed implant models may serve as basis for patient specific, custom-made implant design using modern manufacturing techniques for example rapid prototyping [15].

Using the example of a female patient (age: 42 years, height: 178 cm, weight: 70 kg, PSI: 2.5) with symptomatic cardiopulmonary compromised pectus excavatum (Fig. 4), the computer-aided surgical planning of chest wall repair with the semi-open Nuss technique, combined with horizontal sternum osteotomy is presented (Fig. 5). Additionally to the 3-D surface scan in standing position (Fig. 1) a preoperative CT scan of the patient in supine position with raised arms was accomplished. By means of specific software the bones and cartilages of the thorax were segmented, three-dimensionally reconstructed and a 3-D virtual model of the skeletal thorax with overlying soft tissue was created (Fig. 5a and b). The degree of the skeletal deformity can now be measured on the virtual 3-D model (Fig. 5b). Specific software functions now enable to virtual osteotomy the deformed thorax and to cut precise slices on instantly the medial and lateral cartilage border (Fig. 5c and d). Already preoperatively the dimension of the osteo- and chondrotomy and the size of excised bony or cartilage segments can be defined (Fig. 5d).

The sternum can be virtually elevated to the desired plane of the rib cage and the resulting alterations can be exactly measured (Fig. 5f). The resulting deformity of the overlying soft tissue (Fig. 5e) is interactively corrected using the above described virtual 3-D morphing software (Fig. 2e). The degree of the surgical correction

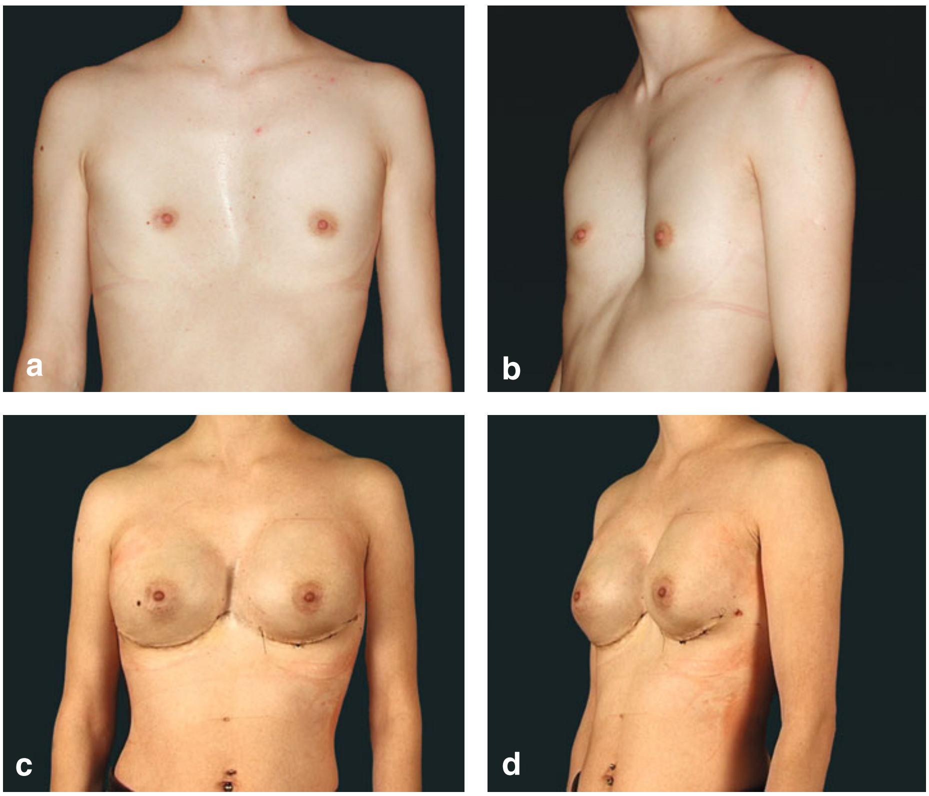

Fig. 6. Preoperative and postoperative 2-D photography of a symptomatic 23-year-old female patient undergoing pectus excavatum correction with Hegemann procedure and combined mamma augmentation: **a** preoperative frontal and **b** preoperative oblique view of the anterior thoracic wall deformity; **c** postoperative frontal and **d** postoperative oblique view of the reconstructed and augmented anterior thoracic wall

is quantified and visualized with the aid of a 3-D comparison and 2-D deviation slices (Fig. 5g–i). The resulting deformity of the thoracic wall and the overlying soft tissue can be defined already preoperatively and also can be used as a supportive and additional device intraoperatively.

13.1.4 Clinical application

Using the example of a female patient (age: 23 years, height: 158 cm, weight: 51 kg, PSI: 2.6) suffering from pectus excavatum deformity with combined mamahypoplasia (Fig. 6) a more sophisticated analysis of the postoperative outcome on the basis of the 3-D surface scan data is performed. The patient underwent thoracic wall reconstruction with Hegemann technique and combined mammary augmentation with breast expander-prosthesis for continuous filling and soft tissue expansion. Beside the 3-D quantification of the pre- and postoperative shape changes, the degree of thoracic wall correction combined with breast expander implantation and the soft tissue expansion is differentiated (Fig. 7).

For 3-D quantification of breast shape changes the pre- and postoperative 3-D scans (Fig. 7a and d) were superimposed and a color-coded 3-D shape difference was accomplished (Fig. 7c). The changes of the breast shape on a horizontal slice (Fig. 7c) and the sternal elevation in a sagittal slice (Fig. 7e) of the pre- versus postoperative superimposed 3-D surface scans can be analyzed. The lateralization of the nipple-areola-complex caused by the thoracic contour correction and soft tissue augmentation is clearly detectable (Fig. 7e). Postoperatively the implanted expander was continuously filled, before and after every expansion a 3-D surface scan of the breast region was performed to document the soft tissue change over time (Fig. 7f). At every expansion step the infiltrated volume was correlated with the breast volume increase and surface skin expansion

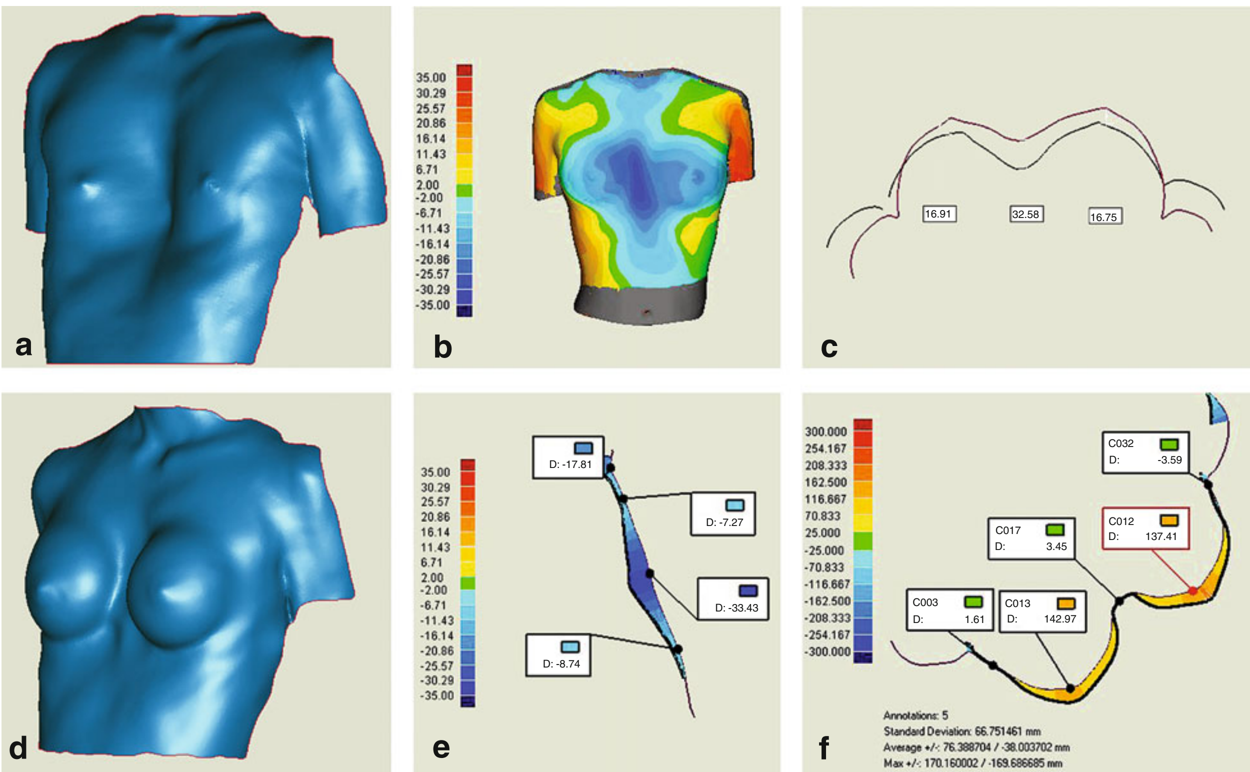

Fig. 7. Quantification of chest wall and breast changes of surgical pectus excavatum correction with simultaneous implantation of breast expander-prosthesis and postoperative soft tissue expansion: **a** preoperative 3-D surface scan; **b** color-coded quantification of 3-D chest wall shape change pre- versus postoperative 3-D scan; **c** color-coded breast shape change after surgical correction before first expansion; **d** postoperative 3-D surface skin after prosthesis expansion; **e** sagittal color-coded 2-D deviation of midsternal contour change pre- versus postoperative; **f** horizontal color-coded 2-D breast shape change after multiple expansions. All measurements in mm

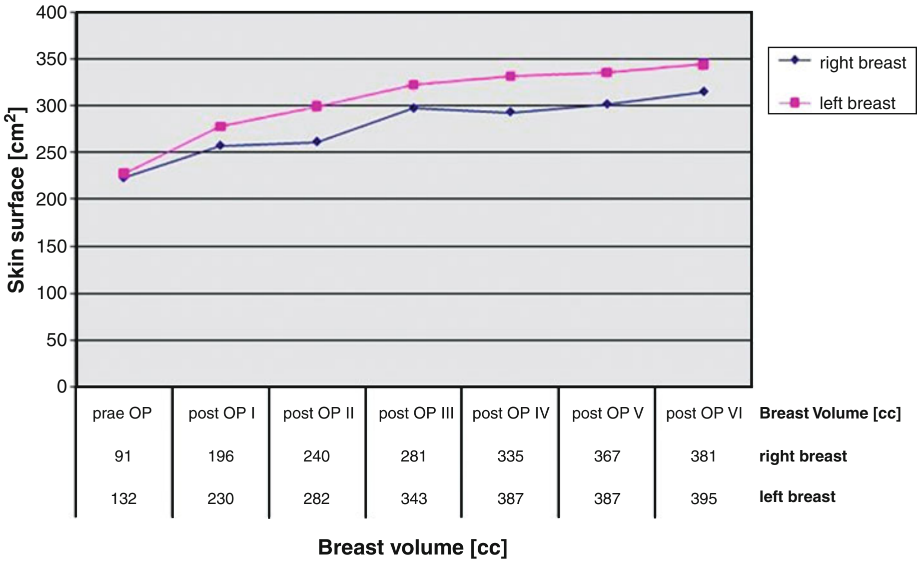

Fig. 8. Quantification of the breast skin surface expansion (cm^2) depending on the breast expander volume (cc) at different post OP expansion dates

according to the breast expander volume (Fig. 8). With the aid of this continuously 3-D quantification a precise and objective analysis of the relation between the breast expander volume and the resulting skin surface expansion is feasible. Beside a precise postoperative documentation over time and a postoperative quality assurance of the surgical outcome, this close interval quantification provides the surgeon with objective data to determine the planning and date of a potential implant exchange.

13.1.5 Conclusion

Particularly with regard to conventionally used 2-D photography the pros of 3-D surface imaging are enormous. Especially the example of computer-aided planning of a custom-made silicone implant using 3-D morphing systems demonstrates the considerable benefit of the 3-D imaging technique. In contrast to the common moulage technique with the patient in supine position, 3-D planning can be performed with the patient standing. With the patient in supine position the overlying soft tissue above the sunken sternum is deformed differently compared to the standing position, in which the true deformity is presented more realistically. However the 3-D intuitive modeling of the surface contour is limited as well. The true skeletal thoracic wall contour on which a designed implant will be placed, is masked by the overlying soft tissue during surface measurement. Therefore efforts have been undertaken to design custom-made implants using CT data taking this soft tissue coverage into account [13]. But this technique is limited by its radiation burden, time and cost intensive assessment and soft tissue change during the assessment (Fig. 9).

Optical 3-D scanning systems allow static 3-D surface geometry assessment. The clinical example of the computer-aided correction of the pectus excavatum repair with the semi-open Nuss procedure with combined osteotomy shows that the required thoracic contour changes and the degree of the performed osteotomy are preoperatively quantifiable and the aspired results can be predicted (Figs. 4 and 5). But the actual 3-D geometric prediction does not take the biomechanical properties of the human tissue into account. A surgeon will surely gain benefit from a tool that enables precise preoperative planning of the size and extent of resection at bone and cartilages, but the resulting interaction between the elevated thoracic bone and the overlying soft tissue cannot yet be simulated perfectly. The solely 3-D interactive modeling without taking biomechanical soft tissue parameters into account provides the surgeon with reasonable intuitive surgical planning information, but delivers only supportive objective measurements and thus the experience of the surgeon is still essential for the result. Therefore one cannot refer to 3-D surface scanning and modeling as surgical simulation. By

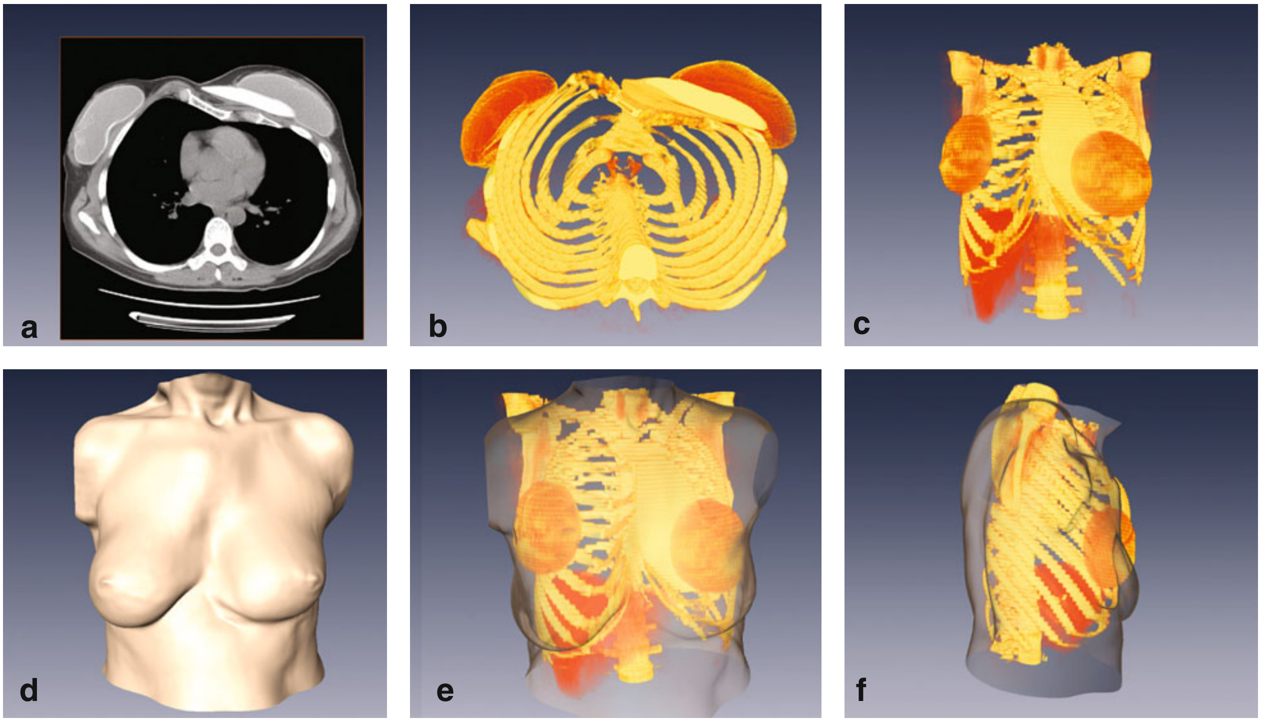

Fig. 9. Patient obtained surgical pectus excavatum correction using thoracic wall silicone implant and breast implants: **a** CT scan in supine position with implant localization; **b** 3-D supine CT data visualization of the implants view from below and **c** view from in front; **d** patients 3-D surface scan in upright position; **e** 3-D visualization of the soft tissue deformation and resulting implant upward localization between 3-D CT data in supine position and superimposed 3-D surface scan data in upright position view from in front and **f** from lateral right side (© CAPS/Brossmann)

definition a virtual simulation presumes faithful description and reproduction of biological processes using mathematical and physical precise models which can be transferred to reality.

To optimize numerically correct simulations it would be necessary to implement physical soft tissue parameters in the existing mathematical models to consider tissue elasticity and stress conditions. The implementation of these factors in the reconstructive surgical planning and to talk about a realistic virtual simulation by definition will be the challenge of future research activities in the field of 3-D imaging. First approaches can be found in the field of finite element method, which allows to mathematically calculate biomechanical processes with the aid of a numerical 3-D simulation model. Efforts to simulate the soft tissue deformation after complex osseous remodeling can be found in cranio-maxillo-facial surgery with very promising results [6]. To improve surgical techniques in the future it will be necessary to combine the results of different acquisition methods (CT/MRI and 3-D surface scans) in multi modal concepts, which contain information regarding tissue consistence and specific characteristics.

As consequence for the future clinical routine a standardized acquisition position and procedure must be maintained. Especially when combining CT/MRI data and 3-D surface scan data sometimes considerable difficulties are encountered. The reason for this problem is the different body position during acquisition. During the CT/MRI patients are in supine position with resulting soft tissue deformities, which causes extreme contour changes in the female breast region compared to the standing position during 3-D surface scanning (Fig. 9). The degree of the position depending soft tissue deformation must be analyzed and simulated before correct biomechanical surgical simulations can take place.

Altogether the advancements of the 3-D technology for surface assessment of the human body surface enter unchartered territory to quantify shape and volume changes of the human body. The currently available optical systems permit adequate precise and accurate 3-D visualization of the human body and on the one hand facilitate the surgeons estimation and planning of the extent of surgical correction of severe and complex defects and on the other hand the communication with the patient regarding potential limitations and objectives of a desired result. Although the research results with 3-D imaging data are very promising, these methods have to be validated with larger patient collectives before introducing it as a routine tool in clinical evaluation. However the empirical and intuitive estimation of an experienced surgeon will never be totally replaced by modern technologies, but such techniques and developments can support teaching or comprehensibility for patients.

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13.2 Special instruments, technical refinements

Anton H. Schwabegger

All measurements which intend to repair a pectus deformity nowadays are aimed to utilize techniques as minimally invasive as possible to avoid growth disturbances during childhood [6] and extensive scarring in adults likewise. Such late complications were ascribed earlier [6, 14] resulting from extensive mobilization of thoracic wall tissue and forced remodeling procedures during childhood. Nowadays many studies are available that deal with large series of experience and long-term follow-up particularly in children with the MIRPE and similar techniques. Since the advent of the MIRPE technique [15, 16] many minor modifications and refinements were developed, also major ones for the application in adults (MOVARPE). To describe all advantageous or potentially also adverse new techniques would definitely exceed the scope of this book.

Herewith exemplarily listed special instruments and refinements shall just show up the progress of development. Even these, among many others, highlighted techniques may be overcome in the near future and replaced by other refinements, which will be elaborated based on creative findings, further long-term experiences, and scientific work worldwide.

13.2.1 Round tunnelizer

In the MIRPE technique initially large clamps were used as a tunnelizer or as an introducer for the intrathoracal and retrosternal manipulation in children [15, 16]. By development of particularly refined instruments for the specialized pectus excavatum repair, a strap like tunnelizer/introducer was designed, with a minor size for children and a major sized and reinforced one for adolescents (Fig. 1a–c). Such reinforced tunnelizers definitely alleviated the passage through the thoracic cavity and as a further advantage due to their robust features they allowed intraoperative remodeling of the anterior thoracic wall by preliminary elevation prior to and sequently alleviating the final transthoracal retrosternal passage of the pectus bar.

By further extensions of the application of the MIRPE technique to athletic adolescent and adult patients, problems with rigidity of the matured skeleton arose. The inelastic adult ribs rather rigorously withstand the introducing forces of metal hardware, thus increased manipulation strength must be applied for their passage leading to shearing forces along the rib edges. Such undue violence with an edged instrument will definitely damage the subcostal neurovascular structures as well as the intercostal muscles with periosteum (Fig. 2a and b). Furthermore, intrathoracally shearing forces with edged hardware retrosternally may damage the internal thoracic vessels with resulting hemorrhage [26]. In order to circumvent such potential complications particularly

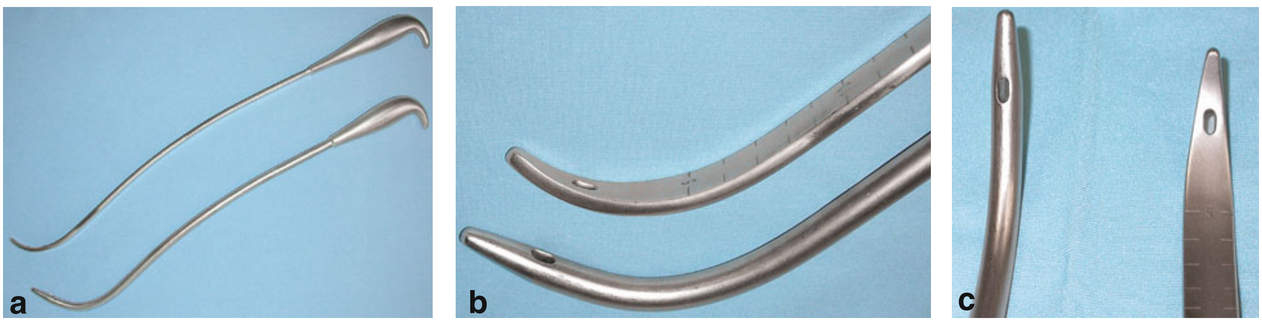

Fig. 1a–c. Comparison of two different types of tunnelizers. The flat one shows edges, which however are slightly rounded but are able to disrupt vessel and nerves at the intercostal or parasternal area. The round one is lesser prone to cause shearing-off effects of tissue from bones during intrathoracic manipulation

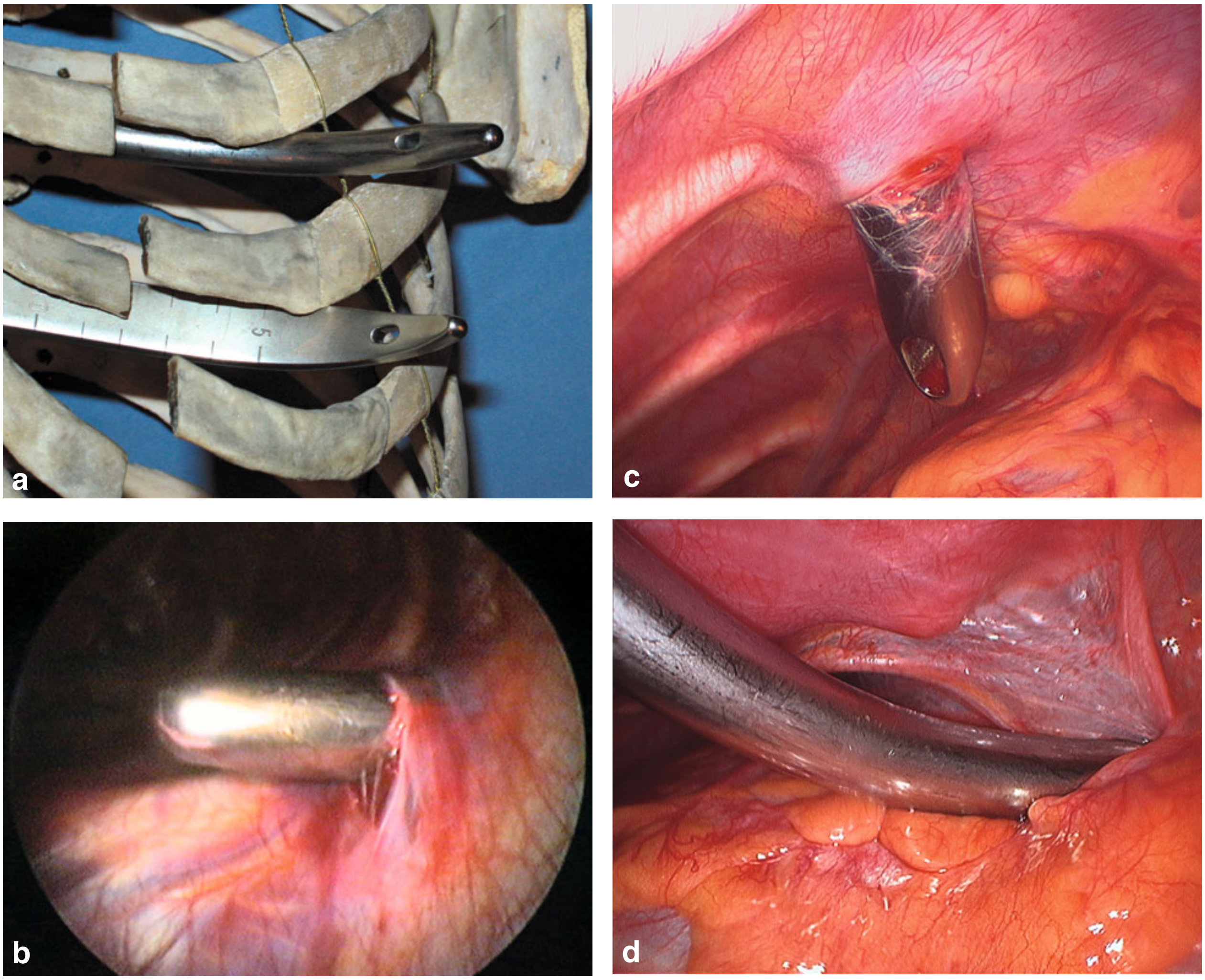

Fig. 2a. Skeletal specimen to depict the beneficial round shape of the rounded tunnelizer (above) versus the edged at one intercostal space below. **b, c** VATS view at the moment of perforation of the tunnelizer into the thoracic wall at the intercostal muscles, the round tip is closely passing the intercostal vessels and nerve without traumatization. The perfectly round tip and round shape allows for blunt perforation thus minimizing any bleeding at the thoracic wall. **c** The round tunnelizer by its advantageous shape avoids damage to the internal thoracic vessels (in the rear left from the tip of the instrument). **d** Dissection of the mediastinal and pericardial fat (below the instrument) from the sternum with gentle fan-shaped movements and loose contact to the posterior surface of the sternum

in adult patients, a rounded dissector was developed. The perfectly round diameter (Fig. 2c–e) only hardly might damage any structures along its passage between ribs retrosternally.

13.2.2 Angled oscillating saw

Such a specially shaped saw (Fig. 3a) is advantageously used for osteotomies with an access along a long subcutaneous tunnel (Fig. 3b) that can be placed at a lower thoracic region, when no space for osteosynthesis manipulation is required. In order to provide a proper survey of the osteosynthesis site furthermore an angled retractor with attached cold light source is used. Such modified retractors, frequently used in breast augmentation surgery may be very hepful in females with surgical acces from the submammary crease (Fig. 3b).

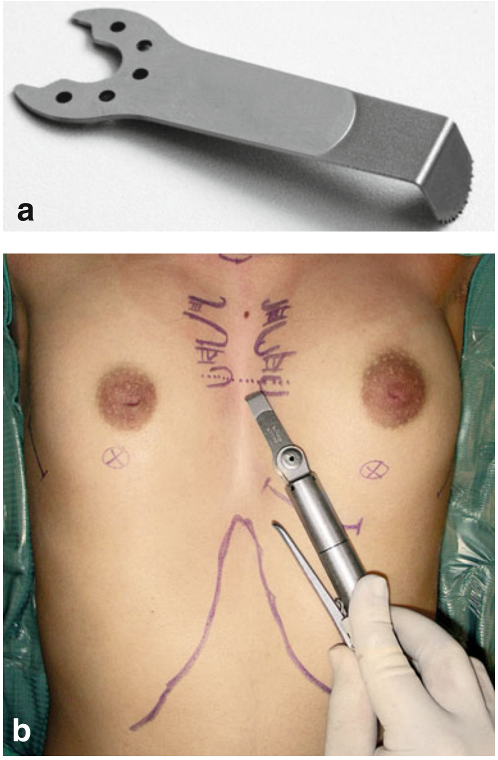

Fig. 3a. Angulated saw blade to be used at an oscillating driller for horizontal incisions at the sternum. **b** Saw instrument held in place to demonstrate the advantageous use of an angulated saw via subcutaneous access from the inframammary crease to the sternotomy site

13.2.3 Extrapleural pectus bar

Schaarschmidt in 2005 described a variant in the retrosternal and intrathoracal passage of the pectus bar. In up to 90% of cases an extrapleural positioning of the pectus bar should be feasible thus reducing the common complications associated with the MIRPE technique, namely pneumothorax, pleural effusion, and pain. By that the routine use of chest tubes was also diminished to a minimum. It may be expected that pleural adhesions ensuing to such extrapleural passage are reduced to zero, thus redo operations or required thoracoscopic interventions due to other reasons are not complicated by intrathoracal scars [21]. However, such manipulations require experience and improved technical skill with endoscopic procedures and prolong the surgery time for several minutes. Its effort on the other hand seems to be justified and worthwhile with respect to the reduction of typical early and late complications of pleural lesions.

13.2.4 Pectus bar fixation

Several refinements with the fixation procedure of the lateral wings of a pectus bar are described, using lateral stabilizers either of metal or of bioabsorbable material [19, 28]. Furthermore several kinds of wires [29, 31], sutures, or threaded alloplastic material are used in different institutions depending on the requirements of fixation forces and personal experiences with different materials. However, no statistical data can describe or propagate any material with superior characteristics and properties above others, with respect to acting as an optimal device for every kind of application. Almost every material so far described and experienced contains a list of advantages and disadvantages (Table 1), on which the performing surgeon has to rely on and select the appropriate one adapted to the individual require-

Table 1: Different characteristics of a variety of fixation devices for the pectus bar wings

Material	Advantage	Disadvantage	
Metal stabilizer	Stable fitting to the pectus bar	Palpable bulk, intra- and perimuscular scarring, difficult to remove	All kind of circumcostal suturing is prone to damage of the intercostal neurovascular bundles; the more rigid the suture the more the probability of vessel and nerve strangulation
Bioabsorbable stabilizer	Stable fitting to the pectus bar, auto-absorption	Palpable bulk, may break, increased wound infection rate, intra- and perimuscular scarring, costs	
PDS® sutures	Strong suture with long-term absorption	Thread knot-ends may be palpable for months	
Mersilene® band	Very strong device	Not absorbable	
Metal Wire	Very rigid fixation to the ribs feasible	May breake and migrate into the thorax	

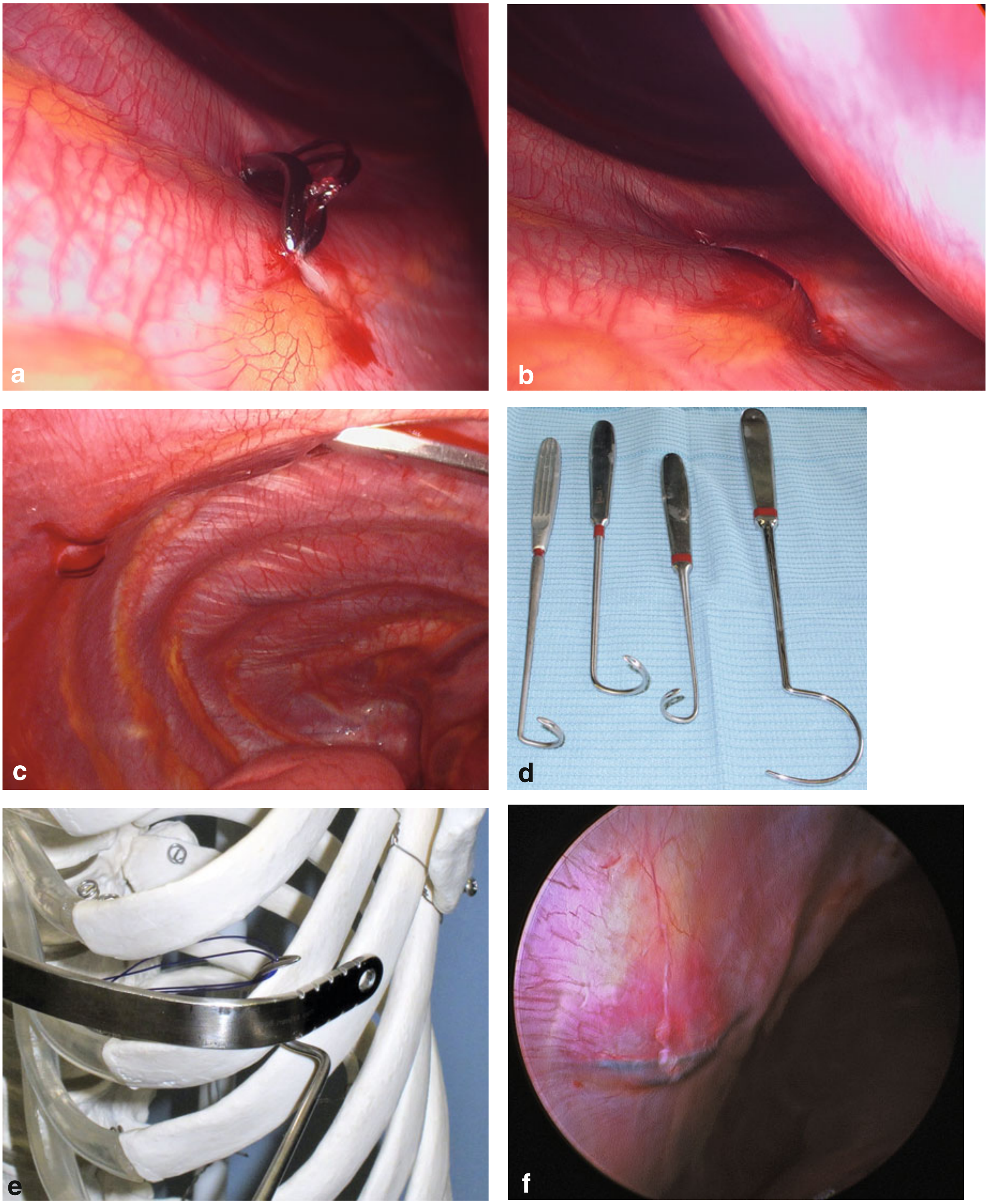

Fig. 4a. VATS view at a PDS threaded Deschamps needle just passing the inner surface of the rib. **b** Completion of circumcostal suture seen at the lateral thoracic wall. **c** Overview within a right thoracic cavity and deflated lung. Notice the pectus bar at the anterior thoracic wall in situ (above), its lateral wing fixed with two circumcostal PDS sutures (left side at picture). **d** Deschamps ligation needles with different shapes for the pectus bar fixation. The angled ones serve for lateral fixation, the straight and large one (right) for parasternal access. **e** Skeletal specimen to depict the course of the Deschamps needle with suture around the rib and attached pectus bar wing. **f** Subpleural course of the Deschamps needle to avoid damage to the pleura thus minimizing complications

ments of the patient. If ever feasible, a three-point fixation [17, 30] in patients beyond childhood, that is an additional fixation at the parasternal region should be performed, which is feasible anyhow, when an additional central surgical acces (MOVARPE) is applied.

Such pericostal or better described as circumcostal fixation with exemplarily PDS sutures may be achieved with several devices available [7, 20, 29, 30]. Ideally the fixation procedure, that is the circumcostal, transmuscular, and endothoracal passage of the threaded instrument is guided and surveyed with the aid of endoscopic view (Fig. 4a–c). A Deschamps ligature device with a blunt tip is available in different shapes (Fig. 4d) for variable accesses to the ribs laterally (Fig. 4e) and in the parasternal region to place the sutures around the ribs. Care herewith has to be taken to avoid lesions of the neurovascular intercostal bundles during the needle passage and only loose knotting is recommended to prevent the intercostal nerve, running within the subcostal groove from strangulation. Demanding some skill, the circumcostal passage may even stay at the extrapleural space (Fig. 4f) thus minimizing pleural adhesions through the surgical trauma or bleedings into the pleural cavity, potentially derived from lesion of the intercostal vessels [21].

13.2.5 Hybrid repair

Modified approaches utilizing the MIRPE technique in combination with minor- or semi-open (MOVARPE) access seem to further establish its right for application in cases with major deformities with sternum malrotation, asymmetry or in adolescent and adult cases with thorax rigidity. The elevation and ensuing alleviated support of deeply sunken sternum in extensive and/or asymmetric funnel chest deformities must be preceeded by multiple relaxing incisions at the sternum and parasternal rib cartilages, occasionally also by wedge resections and chondrectomies. Minor cutaneous incisions at the preseternal region may accompany the incisions needed for the MIRPE access, but compared with conventional open techniques the hybrid access still is of minor invasiveness with reduced pain due to reduction of the forces needed to overcome the rigidity of an exemplarily athletic thorax or of tall and bodily matured patients (Chapter 6.4).

As a refinement of such a hybrid access in the pectus excavatum repair, multiple relaxing- or so-called kerf incisions were applied by Al-Assiri et al. [1] from inside the thorax under endoscopic view in order to relax the anterior thoracic wall prior to the pectus bar implantation maneuver. The outcomes were not different in terms of aesthetics or function, but the intraoperative view during the intrathoracal maneuvers was definitely improved, thus providing increased security to avoid lesions in thoracal organs. In addition, a relaxed anterior thoracic wall will alleviate the retrosternal passage of instruments and pectus bar thus reducing the necessity of undue forces during their implantation.

13.2.6 Absorbable plates and screws

Although widely used in craniofacial surgery [9] but to a limited extent in the throacic wall repair [4, 11–13, 25], they still do not provide with the same stability as does metal hardware with respect to bending forces, which have to resist against forces after anterior chest wall remodeling. The resistance to longitudinal strain however is similar to those of metal mini-plates with the major advantage of complete absorption after 1 year. On the other hand, such elastic properties follow the micromovements at the sternum osteosynthesis during respiration, thus behave like a dynamized nailing concept with resulting stimulus to form callus for fracture healing by secondary intention. Usually with an oblique and subcutaneous access a perfectly tight osteosynthesis enabling primary fracture healing in the sternum region is not feasible due to limited survey and limited space for surgical maneuvers in the pectus deformities repair with predomonantly aesthetic claim. The loss of resistance from slowly proceeding absorption takes place far beyond the time of completed biological fracture healing [9, 11], so that temporary stabilization with such material suffices the requirements of the remodeling repair at the sternum. However, plates and screws will not at all provide stability at strained cartilages and should not be utilized at the ribs instead of ideally applied strong reefing sutures.

13.2.7 Absorbable lateral stabilizer

Lateral stabilizers with different styles (Fig. 3a in Chapter 6.9) were and still are widely used as an adjunct to avoid tilting of the pectus bar. In order to circumvent cumbersome maneuvers and tissue dissection for the purpose of pectus bar removal (Chapter 6.9), absorbable stabilizers have been developed. Such absorbable devices represent an interesting development in order to reduce the surgical trauma and life quality during and after a pectus bar removal procedure. However, they are prone to show more complications in terms of device fracture or wound infection at the site of implantation compared with the metal ones [19, 28]. On the other hand, several authors already abandoned the use of lateral stabilizers at all, since they preferably rely on measurements using minor volumes of foreign material but apparently improved statics by three point fixations with sutures, wires, or other threads to successfully avoid tilting of the pectus bar [7, 29].

13.2.8 Klobe's vaccum bell

When a patient due to several reasons decides against a pectus bar suspension but in favor for the conventional technique according to Ravitch in the repair of pectus excavatum, one could utilize the vacuum bell after Klobe (Chapter 6.8) as an additional supporting device. Such a measurement starting several days postoperatively may maintain the position of the surgically elevated sternum, which by the conventional technique is held in place only by sutures and eventually osteo-

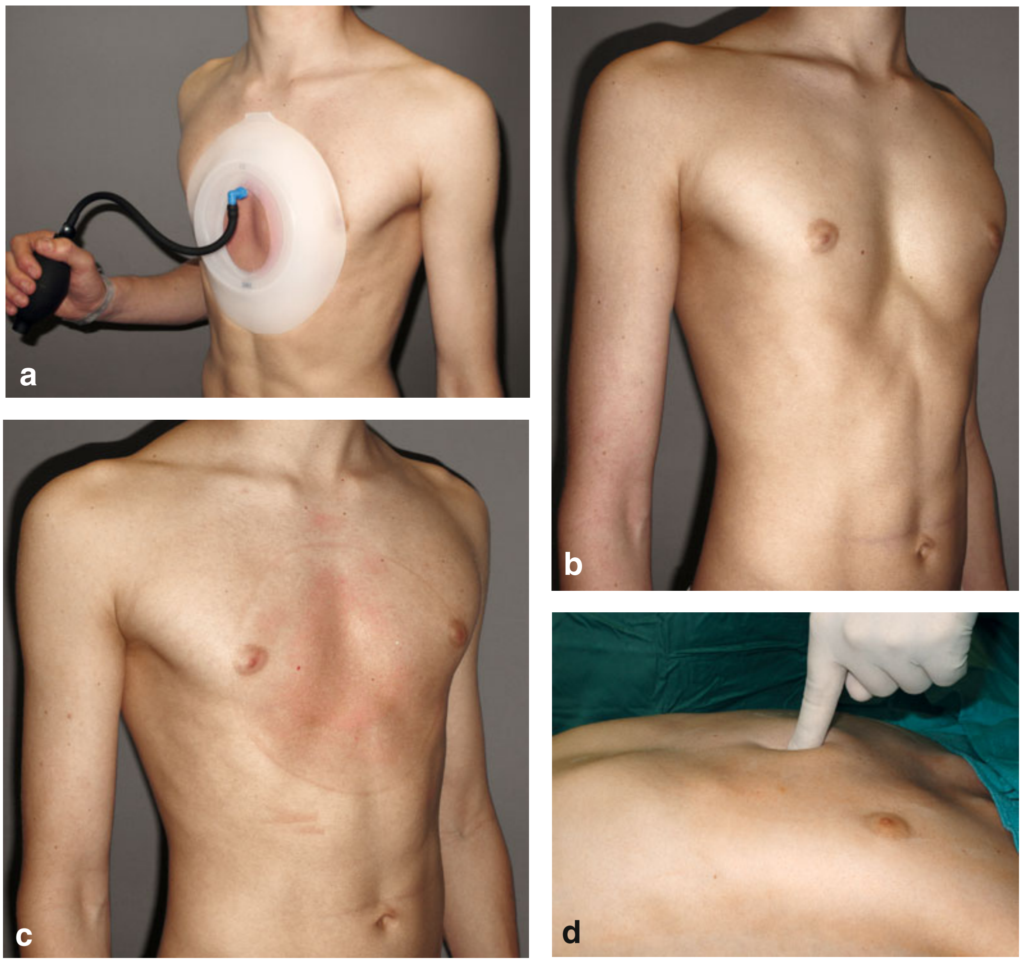

Fig. 5a. Vacuum bell after Klobe is applied to enhance the volume of the soft tissue at the presternal region during a short-term period of application. It may also be used as an intraoperative adjunct to elevate the sternum during and for the purpose to alleviate surgical access with retrosternal passage. **b** and **c** Pretreatment situation of a moderate pectus excavatum deformity (**b**). Situation with temporary tissue augmentation achieved only by several single applications of the vacuum bell (**c**). **d** Intraoperative situs, the anterior thoracic wall is pretreated with the vacuum bell for 3 weeks to moderately elevate the sternum as an assistance to alleviate the retrosternal passage of instruments and pectus bar during the MIRPE procedure. Notice the additional side effect of tissue augmentation by edema, demonstrated by the sunken finger, in fact prepared and suitable for a lipofilling procedure. However, the patient with this pretreatment successfully underwent MIRPE

synthetic plates. Through the negative pressure the natural forces to regain the original shape of a sunken sternum might be overcome to stabilize the long-term result therewith (Fig. 5a).

There is however no such application yet described in the available literature. The original purpose of the application of the vacuum bell to an untouched and surgically untreated thoracic wall is to achieve an elevation of the sunken sternum to ventral, overcoming the naturally present forces of disfigured sternum and cartilages [2, 5, 23]. It would be plausible and understandable therefore that a thoracic wall surgically weakened by osteochondrotomies can be held in position or, as an aesthetic advantage, can even be elevated slightly out from the surgically feasible level of elevation. This maneuver by such a negative pressure device therefore appears much easier then than without surgery to achieve a permanently pleasing result. Certainly the compliance of the patient in the postoperative weeks must be ensured herewith. The duration of such an ancillary treatment would be 2–3 months until a stable consolidation of the cartilages and reefed periosteal tubes as well as the osteosynthesis is achieved.

The vacuum bell applies to a technique that is used in the breast augmentation also, named the BRAVA technique. Herewith the negative pressure applied to underdeveloped female breasts is used to augment their volume gradually and over the duration of several months [24, 27]. A recent refinement consists therein that preexpansion of breast tissue by negative pressure just for several weeks leads to volume augmentation by edema and enhancement of vascularization. Into such altered tissue lipofilling is then applied in order to permanently achieve tissue augmentation by improved conditions for the implanted lipocytes to survive. However, such reports are singular yet and series with more patients treated are not available. From a logical point such an enhancement of vascularization and subcutaneous volume by negative pressure within a funnel chest depression (Fig. 5b–d) as a priming for ensuing lipofilling might be a valuable tool to improve the late results of lipofilling in the delicate presternal region.

13.2.9 Endoscopic and thoracoscopic repair in pectus carinatum

This seems to be a promising development in terms of reduction of major presternal scars and pains that result from the invasiveness of the common open access [10, 22]. Partial resection of abundant length of rib cartilages seems to be feasible from a thoracoscopic approach in unilateral pectus carinatum deformities [10]. If the sternum is involved, the technique after Schaarschmidt, using a subpectoral but endoscopic access may be applied to reduce invasiveness and postoperative morbidity compared to a conventional open access.

Hock in 2005 used the MIRPE technique in a reverse manner to repair pectus carinatum deformities in a small series with convincing results so far [8]. The depression of the protrusion of sternum is managed by implantation of a pectus bar above the sternum and is supported by intrathoracal "sublay" of the pectus bar wings bilaterally along the thoracic wall flanks, which are elevated therewith.

However, these preliminary series of minimally invasive endoscopic pectus carinatum repair so far has not yet provided long-term results, furthermore ensuing studies are not available.

13.2.10 Lipofilling

Already described in Chapter 6.6.2 this topic is new and so far only preliminary experiences and publications exist in the application to thoracic wall deformities [18]. The long-term results in the application at other than pectus anomaly sites are promising and further development is still proceeding. One major issue for the application at the central sternal region is the fact that only a thin subcutaneous layer exists with low vascularization, into which lipotransfer may be applied. The relatively strong adhesion of skin to the sternum impedes the injection of fat and its survival, since pressure is well known to inhibit ingrowth of transplanted fat. It might therefore be wise to apply lipofilling predominantly at lateral thoracic wall depressions or in combination with vacuum bell application in advance [3].

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Final conclusions

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The comparison of own techniques and results with those from the available literature turns out to develop as an endeavor, because different authors rely on different measurement methods concerning the objective and subjective outcome. Furthermore publishing only own results usually tends to propagate the own methods and techniques, frequently based on modifications of prior developments.

A harmonization of measurement tools and techniques based on evidence thus is desirable, because only therewith comparability in the context of multi-centre studies with sufficient numbers of cases treated would become feasible.

Based on our experiences lasting for more than one decade now in utilizing and developing especially different strategies of surgical and non-surgical treatment, the algorithms of indication setting and patient selection have been revised, taking into consideration all available alternative methods and possible complications. The main conclusion for algorithms in patient selection is to very thoroughly evaluate and even recommend available alternatives before utilizing a method that eventually only meets available surgical skill and a pure aesthetic desire of a patient.

However, conclusive studies are still lacking due to variable surgical expertize or inhomogenous patient collectives, thus the proper selection of the most appropriate techniques for a multitude of indications sometimes remains cumbersome. Only based on such studies and particularly those concerning the long-term follow-up, meaningful techniques will successfully be comparable to experimental interventions and then will survive and overtop the lesser suitable ones.

The contents of this book represent the personal experiences of several contributiong authors, complimented with earlier and recent publications about pioneer work, erroneous developments, and new findings. It is however, and due to the amazing fact of a tremendous increase of interest and emphasis in dealing with anterior chest wall deformities since two decades now, almost impossible to present a complete survey over all therapeutic modalities, even if available via papers or electronic publications. Authors or physicians therefore are kindly requested to accept our apologies, if their modifications, recent developments, or former contributions to the development within diagnostics and therapy were not cited due to the above-mentioned reasons.

Anton H. Schwabegger

Syllabus

Bender	instrument to shape the pectus bar
Chondrogladiolar keel	deformity concerning the lower part of the sternum
Chondromanubrial keel	deformity concerning the upper part of the sternum
Coping (psychology)	is the process of managing taxing circumstances, expending effort to solve personal and interpersonal problems, and seeking to master, minimize, reduce, or tolerate stress or conflict
CT	Computed Tomography, ideal for measurement of internal thoracic diameters and three-dimensional depiction of skeletal structures
custom-made silicone implant	individually shaped implant, derived from a moulage kitt that fits to the patient's deformity
DIEP-flap	deep inferior epigastric perforator microvascular free flap, similar to a TRAM-flap but without muscle, for tissue replacement or augmentation, predominantly for breast reconstruction
Deformation	morphological anomaly as a consequence of a mechanical force
Disruption	morphological anomaly as a consequence of an external agent
Endoscopically assisted	operation with the aid of and secured by an endoscope
FCI-flap	free microvascular FascioCutaneous Infragluteal flap, for several indications of autologous tissue transplantation
Flipper	instrument to turn the pectus bar
ICD-10	the *International Statistical Classification of Diseases and Related Health Problems* 10th Revision, is a coding of diseases and signs, symptoms, abnormal findings, complaints, social circumstances, and external causes of injury or diseases, as classified by the World Health Organization (WHO)
IMAP	Internal Mammary (Thoracic) Artery Perforator
MDCT	Multi-Detector row Computed Tomography
MEDLINE	online medical literature database
MIRPE	Minimally Invasive Repair of Pectus Excavatum
MOVARPE	Minor Open Videoendoscopically Assisted Repair of Pectus Excavatum, a hybrid technique of minor open technique combined with the MIRPE, especially applied in major deformities and adults
Moulage	self hardening (vulcanizing) two-component plastic material or plaster cast
MPRs	multi-planar reformatted images
NAC	nipple-areola complex
Nuss	Donald Nuss, M.B., Ch.B., Pediatric Surgeon
OMIM	Online Mendelian Inheritance in Man (http://www.ncbi.nlm.nih.gov/omim/)
PACS	Picture Archiving and Communication System, a digital medical management, communication, review, and distribution system
Pectus arcuatum	mixed deformity with protrusion and depression of the anterior thoracic wall
Pectus bar	metal strut, individually shaped for support of sunken sternum and rib cartilages
Pectus arcuatum	mixed deformity with combined protrusion and depression of the anterior thoracic wall

Pectus carinatum	keel chest, protrusion of the anterior thoracic wall
Pectus excavatum	funnel chest, depression of the anterior thoracic wall
Pectus (severity) index (PSI)	also called Haller-index, ratio between largest transversal internal thoracic diameter (a) and shortest sagittal vertebro-sternal distance (b) at deepest inclination of the sternum (PSI = a/b) in a CT scan
PDS	Polydioxanon, polymerized glactin, long-term bioabsorbable sutures
Penetrance	proportion of patients with a clinical phenotype out of a group of mutation carriers
PEEP	*Positive End-Exspiratory Pressure*, raised respiratory pressure in order to detect lacerations of the pleura
Platythorax	flat chest with reduced sagittal diameter along the whole cross section of the thorax
Pouter pigeon breast	*protruding* premature ossification at the angle of Louis, the junction zone between manubrium and corpus sterni, frequently associated with congenital heart defects
PWC	physical working capacity, measurement tool for the fitness
RTV 382	two componential silicone, vulcanizing at room temperature
RSTL	relaxed skin tension lines
SIEA	Superficial Inferior Epigastric Artery based microvascular free flap for tissue replacement or augmentation, predominantly for breast reconstruction
Stabilizer	metal plate fixed to the pectus bar to enhance stability and withstand dislocation
Syndrome	pattern of anomalies or malformations as a direct consequence of one common cause
Tabotamp™	bioabsorbable hemostyptic and antimicrobial fabric mesh made of methylic-cellulose
TMG-flap	Transverse Myocutaneous Gracilis free flap for tissue replacement or augmentation, predominantly for breast reconstruction
TRAM-flap	Transverse Rectus Abdominis (Musculocutaneous) free or pedicled flap for tissue replacement or augmentation, predominantly for breast reconstruction
Tunnellizer	also called introducer, instrument to create a tunnel and guide the pectus bar through the thorax cavity
VATS	Video-Assisted Thoracoscopic Surgery
Voxel	Volumetric pixel, used in medical imaging to depict three-dimensional density values of e.g. computerized tomographic scans
VR	Volume Rendering CT mode for three-dimensional depiction of various tissues/ structures, especially cartilages and vessels
VRAM-flap	Vertical Rectus Abdominis (Musculocutaneous) free or pedicled flap for tissue replacement or augmentation, predominantly for breast reconstruction

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