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-

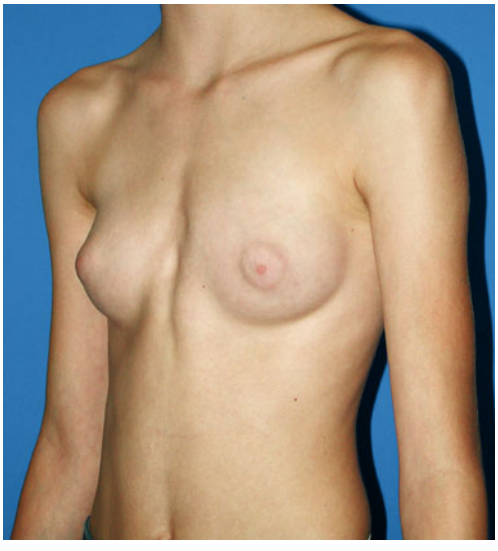

Fig. 1. 15-Year-old girl with isolated rib humps parasternally at the 5th and 6th rib at the left side and a minor neglectable hump at the right side

A. H. Schwabegger (ed.), *Congenital Thoracic Wall Deformities*

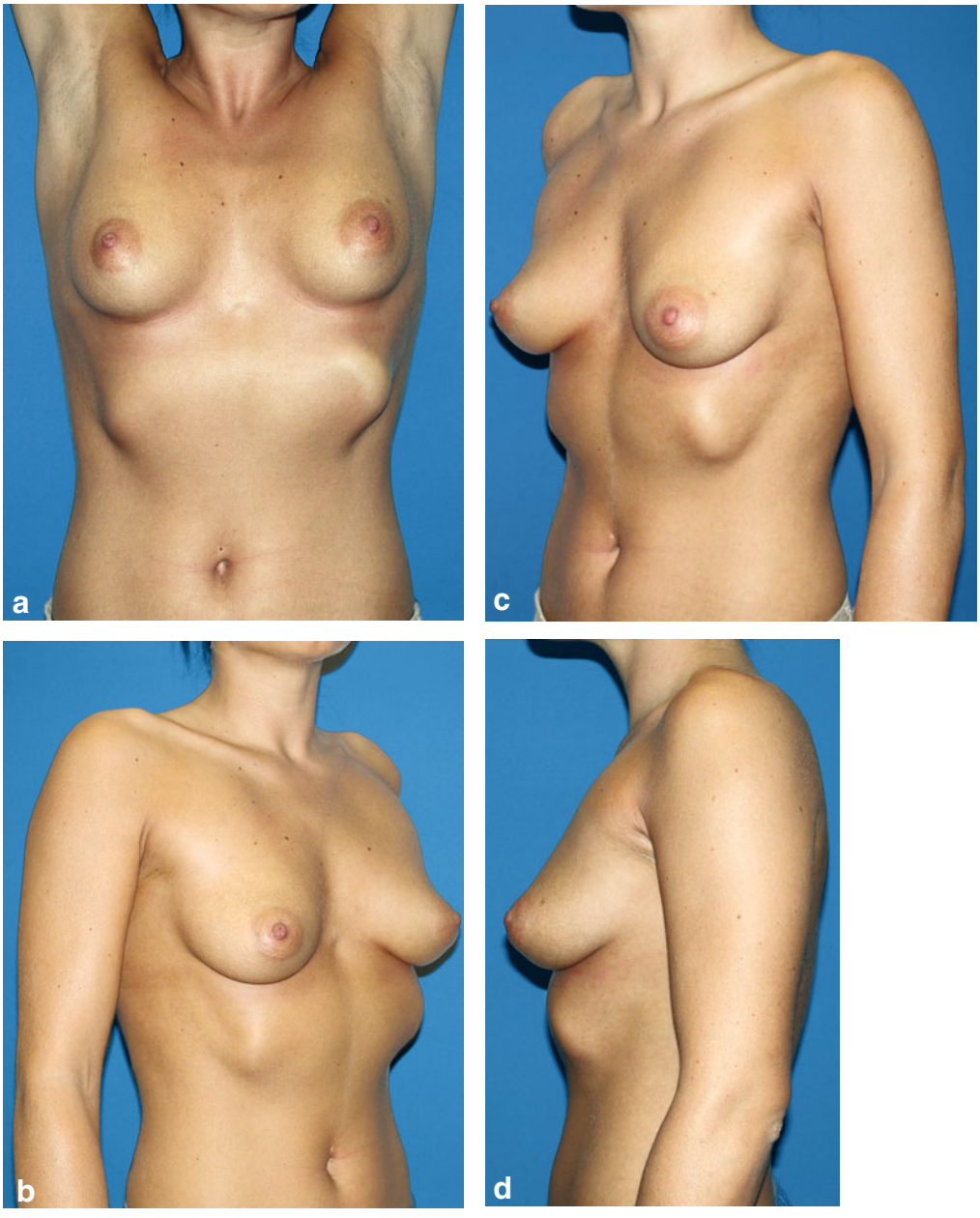

Fig. 2a–d. Preoperative situs in a 24-year-old female with bilateral rib hump deformities prominent at the lower rib arches

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.

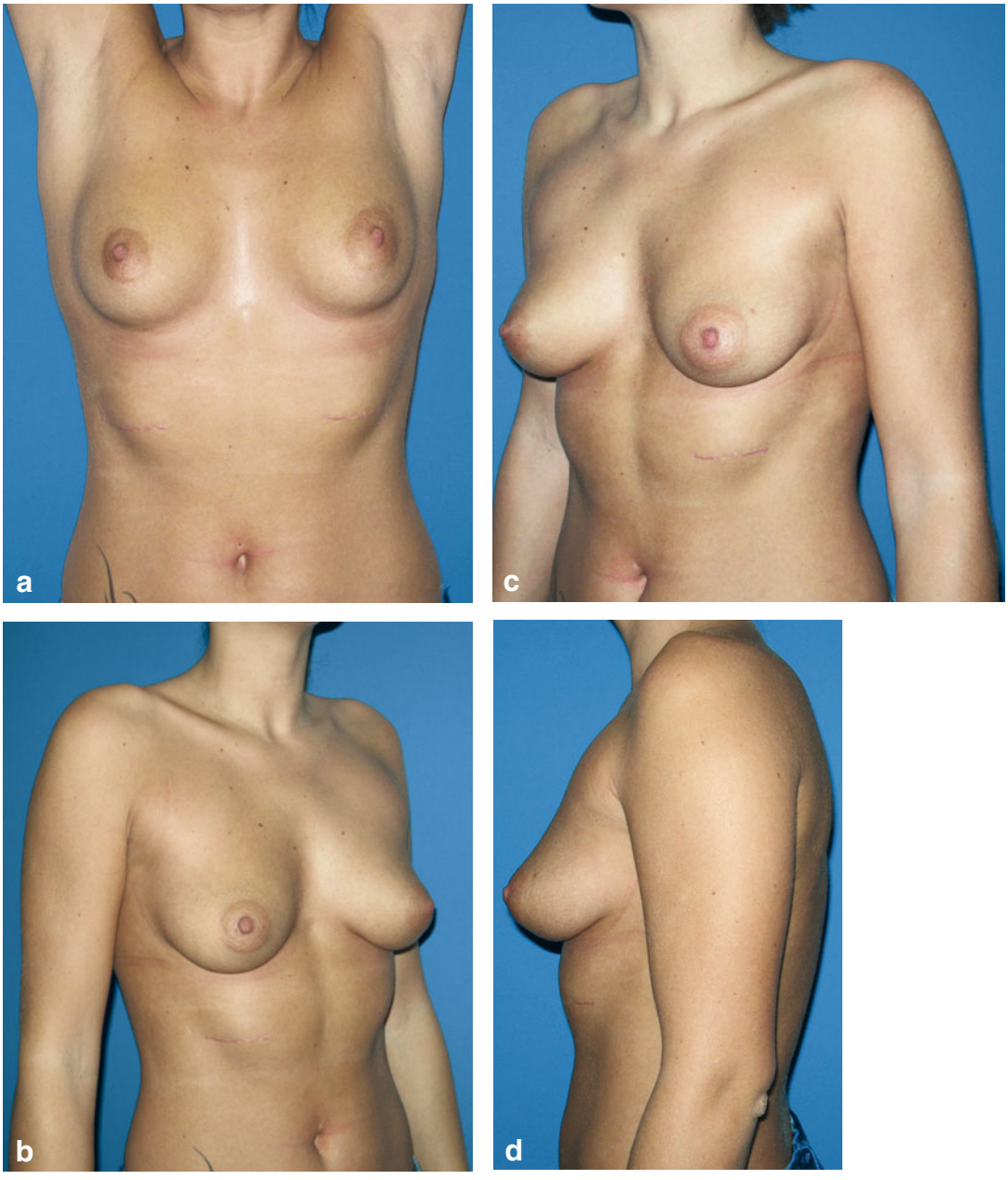

Fig. 3a–d. Same patient 6 months after surgery with resection of the chondo-osseous junction zone of the 7th to 9th rib bilaterally. Skin incisions were set exactly along the RSTL

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.

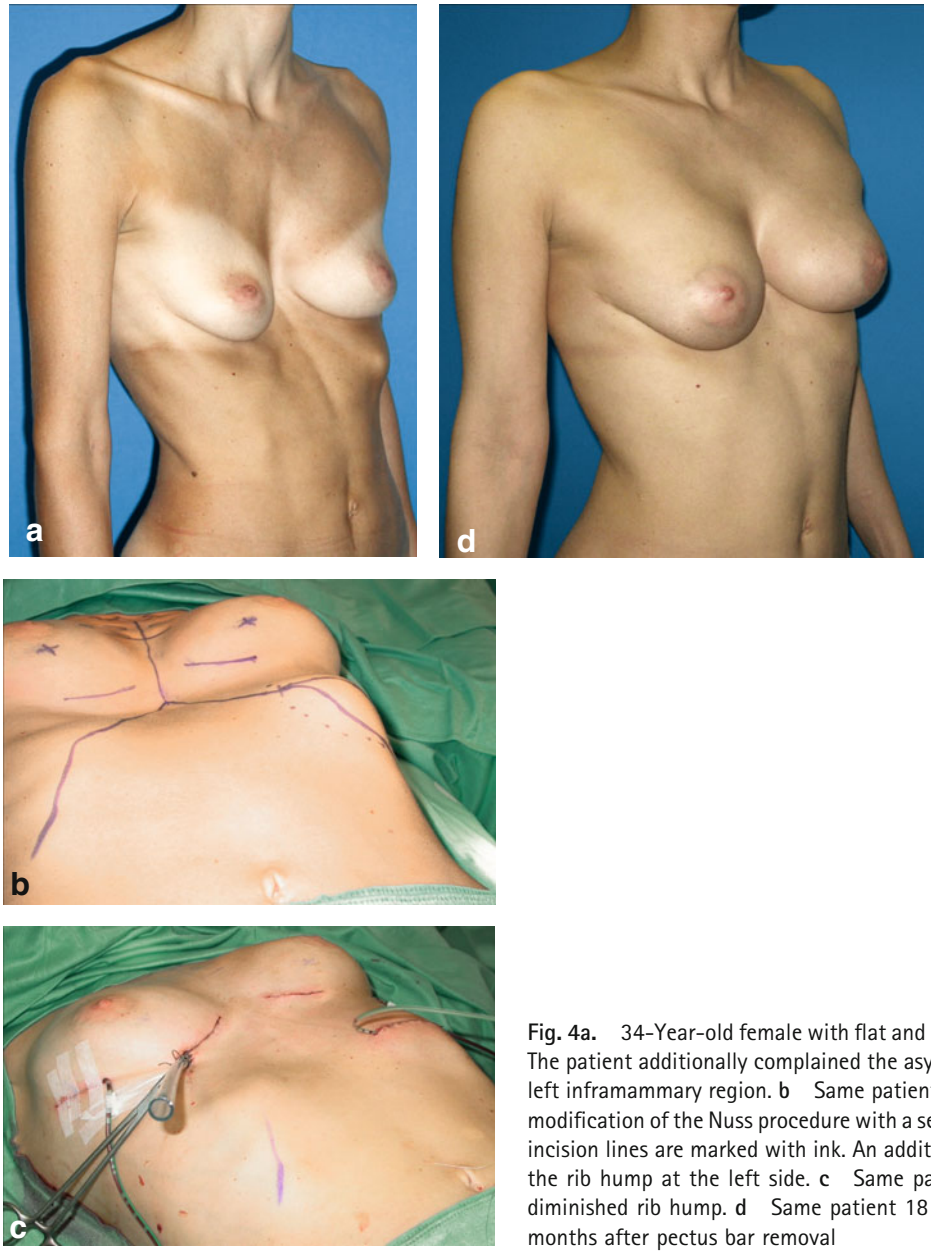

Fig. 4a. 34-Year-old female with flat and asymmetric funnel chest deformity. The patient additionally complained the asymmetric rib hump deformity at the left inframammary region. **b** Same patient intraoperatively, she underwent a modification of the Nuss procedure with a semi-open approach (MOVARPE), 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

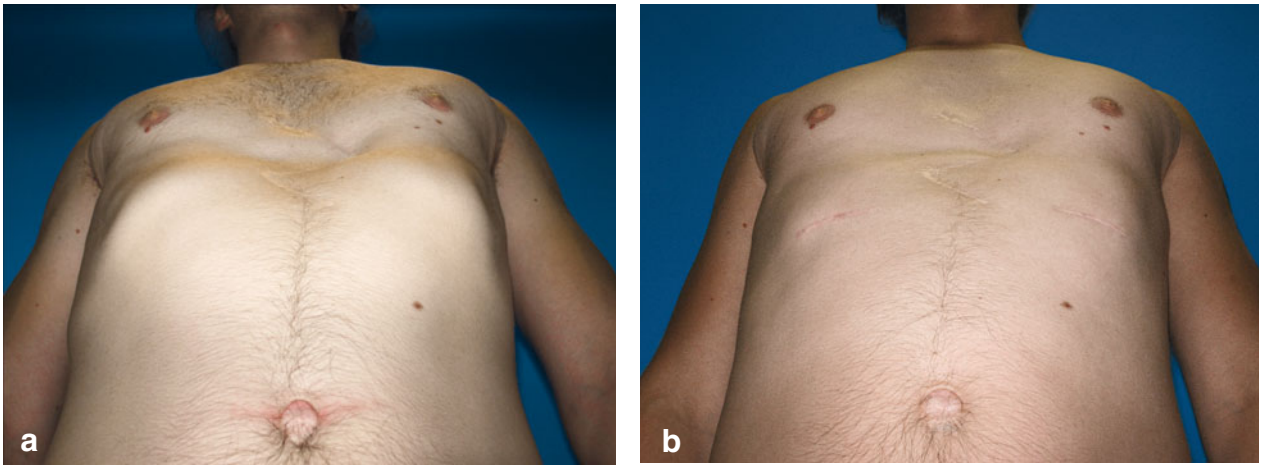

Fig. 5a. 28-Year-old male, 8 years after pectus excavatum correction using the microvascular sternum turn-over technique. Patient complained rib humps at the lower thoracic aperture and minor recidivation of the funnel depression. **b** Same patient 7 months after bilateral resection of prominent rib cartilages and bones. The cartilages were cut to chips and served as a subcutaneous augmentation of the residual funnel deformity presternally

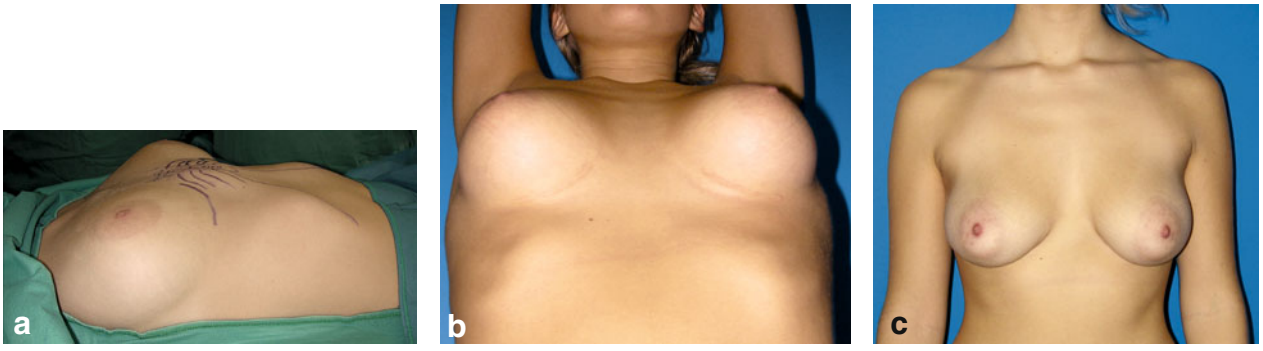

Fig. 6a. 19-Year-old female with bilaterally developed rib humps, representing a minor form of keel chest deformity. Intraoperative situs depicting the planned incisions along the inframammary crease marked with ink. **b** Same patient 1 year after corrective surgery with minor visibility of the scars along the inframammary crease, only visible from beneath and arms elevated. **c** Same patient with well-hidden scars

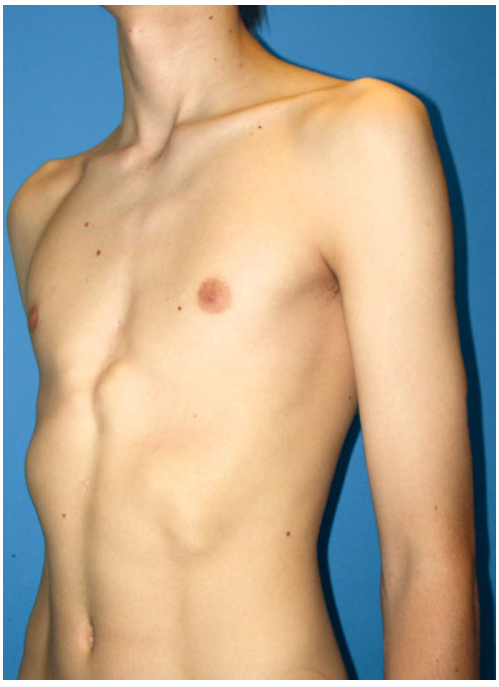

Fig. 7. Isolated minor rib hump deformity at the xiphoid region parasternally left, resulting as a partial recidivation of rib cartilage growth, 1 year after pectus cartinatum surgery

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.

References

[1] Al-Assiri A, Kravarusic D, Wong V, Dicken B, Milbrandt K, Sigalet DL (2009) Operative innovation to the “Nuss” procedure for pectus excavatum: operative and functional effects. J Pediatr Surg 44: 888–892

[2] Hannam S, Greenough A, Karani JB (2000) Rib abnormalities arising before and after birth. Eur J Pediatr 159:264–267

[3] Karami M, Ilharreborde B, Morel E, Fitoussi F, Penneçot GF, Mazda K (2007) Video-assisted thoracoscopic surgery (VATS) for the treatment of scoliotic rib hump deformity. Eur Spine J 16:1373–1377

[4] Kim S, Idowu O (2009) Minimally invasive thoracoscopic repair of unilateral pectus carinatum. J Pediatr Surg 44:471–474

[5] Lieberman IH, Kuzhupilly RR, Reinhardt MK, Davros WJ (2001) Three-dimensional computed tomographic volume rendering techniques in endoscopic thoracoplasty. Spine J 1:390–394

[6] Mummaneni PV, Sasso RC (2005) Minimally invasive, endoscopic, internal thoracoplasty for the treatment of scoliotic rib hump deformity: technical note. Neurosurgery 56(ONS, Suppl. 2):444

[7] Ohara K, Nakamura K, Ohta E (1997) Chest wall deformities and thoracic scoliosis after costal cartilage graft harvesting. Plast Reconstr Surg 99:1030–1036

[8] Ravitch MM (1960) The operative correction of pectus carinatum (pigeon breast). Ann Surg 151:705–714

[9] Robicsek F, Fokin A (1999) Surgical correction of pectus excavatum and carinatum. J Cardiovasc Surg 40: 725–731

[10] Schwabegger AH, Harpf C Ninkovic M, Rieger M (2002) Technical refinements in planning and surgical therapy of pectus carinatum. Chirurg 73:1191–1196

[11] Schwabegger AH, Jeschke J, Schuetz T, Del Frari B (2008) Refinements in pectus carinatum corrections: the pectoralis muscle split technique. J Pediatr Surg 43:771–774

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

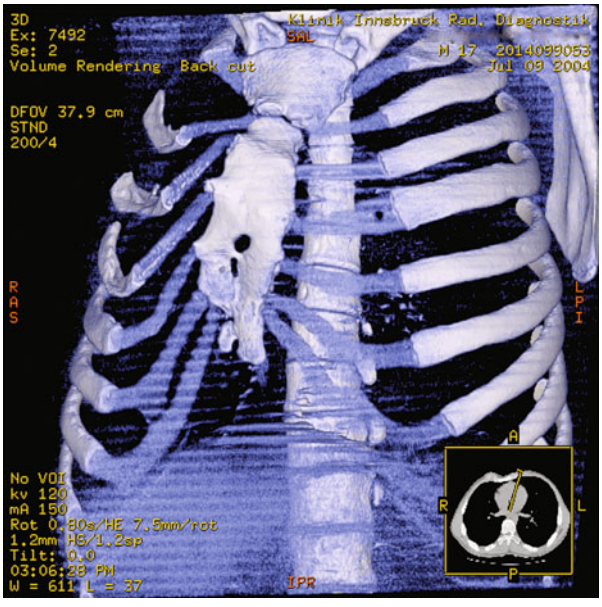

Fig. 1. Volume rendering 3D CT scan of an 18-year old male with major pectus arcuatum deformity and minor cleft sternum deformity

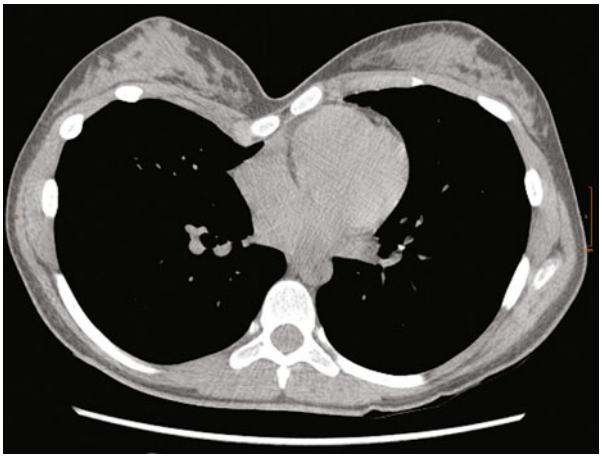

Fig. 2. A 16-year-old female with a sternum foramen associated with a pectus excavatum deformity, predominately complaining the funnel deformity but also the palpable bony deficit at the sternum midline

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.

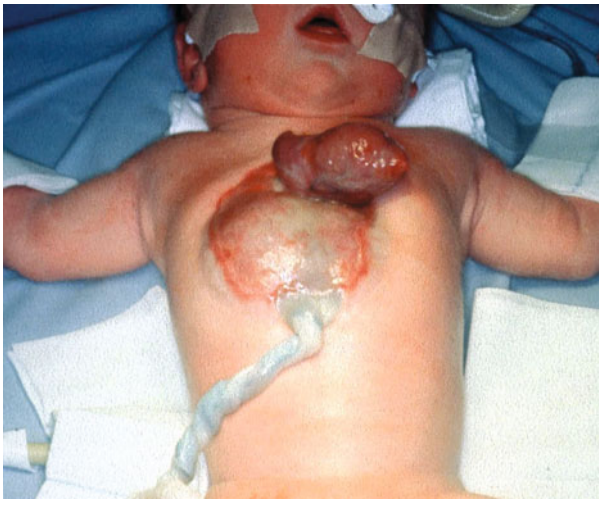

Fig. 3. Newborn with thoracoabdominal ectopia cordis (Picture with courtesy of Prof. J. Hager MD, Department of Pediatric Surgery, Medical University Innsbruck.)

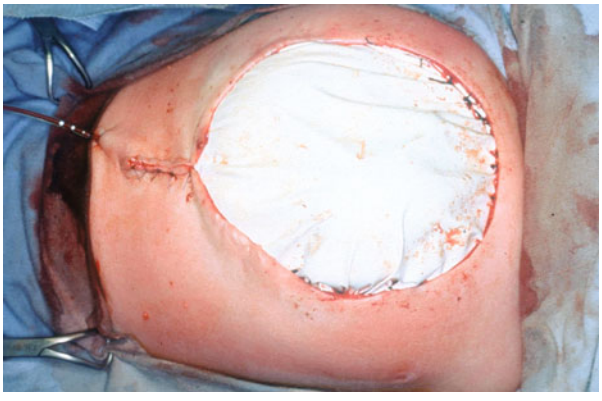

Fig. 4. Same patient as in Fig. 3, staged approach with sequential redressing of exteriorized organs by alloplastic material as a prearrangement for further defect closure. Despite absence of any compression but due to complex and severe cardiac malformations with subsequent cardiac failure the patient 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.

References

[1] Acastello E, Majluf R, Garrido P, Barbosa LM, Peredo A (2003) Sternal cleft: a surgical opportunity. J Pediatr Surg 38:178–183
[2] Alphonso N, Venugopal PS, Deshpande R, Anderson D (2003) Complete thoracic ectopia cordis. Eur J Cardiothorac Surg 23:426–428
[3] Burton JF (1947) Method of correction of ectopia cordis. Arch Surg 54:79–84
[4] Cantrell JR, Haller JA, Ravitch MM (1958) A syndrome of congenital defects involving the abdominal wall, sternum, diaphragm, pericardium, and heart. Surg Gynecol Obstet 107:602–614
[5] De Campos Jr, Filomeno LT, Fernandez A, Ruiz RL, Minamoto H, Werebe Ede C, Jatene FB (1998) Repair of congenital sternal clefts in infants and adolescents. Ann Thorac Surg 66:1151–1154
[6] Dobell ARC, Williams HB, Long RW (1982) Staged repair of ectopia cordis. J Petiatr Surg 17:353–358
[7] Domini M, Cupaioli M, Rossi F, Fakhro A, Aquino A, Chiesa PL (2000) Bifid sternum: neonatal surgical treatment. Ann Thorac Surg 69:267–269
[8] Falcao JL, Falcao SN, Sawicki WC, Liberatori AW, Lopes AC (2000) Cantrell syndrome. Case report of an adult. Arq Bras Cardiol 75:323–328
[9] Greenberg BM, Becker JM, Pletcher BA (1991) Congenital bifid sternum: repair in early infancy and literature review. Plast Reconstr Surg 88:886–889
[10] Haller JA Jr, Colombani PM, Humphries CT, Azizkhan RG, Loughlin GM (1996) Chest wall constriction after too extensive and too early operations for pectus excavatum. Ann Thorac Surg 61:1618–1624
[11] Hazari A, Mercer NS, Pawade A, Hayes AM (1998) Superior sternal cleft: construction with titanium plate. Plast Reconstr Surg 101:167–170
[12] Hill CA, Argenta LC, Hines M (2007) Superior sternal cleft repair using autologous rib grafts in an infant with complex congenital heart disease. Ann Thorac Surg 84:673–674
[13] Ley EJ, Roth JJ, Kim KA, Vincent VR, Muenchow SK, Wells WJ, Downey SE (2004) Successful repair of ectopia cordis using alloplastic materials: 10-year follow-up. Plast Reconstr Surg 114:1519–1522
[14] Luthra S, Dhaliwal RS, Singh H (2007) Sternal cleft – a natural absurdity or a surgical opportunity. J Pediatr Surg 42:582–584
[15] Mathes SJ, Seyfer AE, Miranda EP (2006) Congenital anomalies of the chest wall. In: Mathes SJ (ed) Plastic surgery, 2nd edn. Saunders Elsevier, Philadelphia, pp 457–537
[16] Milanez de Campos JR, Das-Neves-Pereira JC, Velhote MCP, Jatene FB (2009) Twenty seven-year experience with sternal cleft repair. Eur J Cardiothorac Surg 35:539–541

[17] Morello M, Quaini E, Nenov G, Pome G (1994) Extrathoracic ectopia cordis. Case report. J Cardiovasc Surg (Torino) 35:511–551
[18] Sabiston DC Jr (1958) The surgical management of congenital bifid sternum with partial ectopia cordis. J Thorac Surg 35:118–122
[19] Shaw WW, Aston SJ, Zide BM (1990) Reconstruction of the trunk. In: McCarthy JG, May JW, Littler JW (eds) Plastic surgery. WB Saunders, Philadelphia, pp 3726–3796
[20] Snyder BJ, Robbins RC, Ramos D (1996) Primary repair of complete sternal cleft with pectoralis major muscle flaps. Ann Thorac Surg 61:983–984
[21] Suri RK, Sharma RK, JHA NK, Sharma BK (1996) Complete congenital sternal cleft in an adult: repair by autogenous tissue. Ann Thorac Surg 63: 918–919
[22] Valla JS, Bechraoui T, Belghith M, Daoud N, Grinda A (1989) Congenital sternal cleft: closed with periosteal graft. Chir Pediatr 30:219–221