

Pearls and Tricks in Pediatric Surgery

Martin Lacher
Shawn D. St. Peter
Augusto Zani
Editors

 Springer

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Foreword by Benno Ure

Dramatic improvements and expansions in the field of pediatric surgery have been achieved during the last decades. As a consequence, many textbooks have been released dealing with established concepts, new technology and refinements of surgery in newborns, children and adolescents. However, numerous aspects concerning indications, details of operations and their advantages and disadvantages are still controversially discussed within the pediatric surgical community.

The present book *Pearls and Tricks in Pediatric Surgery* adds essential information to this discussion using an exceptional format. Experts answer to questions and thus present their opinion on nearly all relevant issues of pediatric surgery. Their subjective answers are not only valuable information. They also demonstrate that a considerable number of recommendations on dealing with a specific pediatric surgical problem are still under discussion.

The list of authors of this book is extensive and somehow reads like a Who is Who in pediatric surgery. These authors meet the need of opinions on pediatric surgical concepts when evidence for decision making is limited. The pediatric surgical community may be grateful to the editors Martin Lacher, Shawn St. Peter and Augusto Zani as well as the authors for their great work. This book will assist pediatric surgical trainees and experienced clinicians in their daily work.

Benno Ure, M.D.
Editor-in-Chief, European Journal
of Pediatric Surgery
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Foreword by George W. Holcomb

The editors have done an excellent job collating a large amount of available knowledge into a concise, easy-to-read book that will be helpful to all who care for the surgical conditions in infants and children. As in previous editions of this book, the title of the book is really a misnomer. In reality, there are few, if any, secrets in the care of our patients. Rather, this book provides up-to-date knowledge from authors who have a great deal of experience and expertise in their topic. Although I feel the book is directed to the more inexperienced caregiver for infants and children, I think all of us can benefit from the information in this book.

The editors are to be congratulated on a wonderful effort!

George W. Holcomb, III MD, MBA
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Foreword by Agostino Pierro

This comprehensive and innovative book contributes to the dissemination of knowledge that is essential to improve and guide the surgical treatment of children. Drs. Lacher, St. Peter and Zani have finalised a book characterized by comprehensiveness and innovation.

Several chapters have been included covering a magnitude of clinical problems highly relevant to the work of pediatric surgeons as well as trainees in surgery, nurses and other caregivers. The editors have obtained important contributions from a multitude of world class experts working in various countries and in different health systems.

The book is innovative as it is based on questions and answers, therefore highlighting practical problems in the surgical care of neonates and children and reporting the options favoured by the experts. Its value is pivotal for the daily care of these children as well as for the training of a new generation of pediatric surgeons.

Pediatric Surgery is a young specialty which has made enormous progress during the last decades. The outcome of the diseases covered in the book should be further improved, and this book can contribute to refining the surgical management of the children affected. *Pearls and Tricks in Pediatric Surgery* will provide a rapid and detailed reference to clinical problems while discussing the way of managing them. I am expecting that this book will produce a positive impact in Pediatric Surgery and will be utilised by many worldwide.

On behalf of the international community of pediatric surgeons, I would like to express gratitude to Drs. Lacher, St. Peter and Zani for having produced such important contribution in Pediatric Surgery.

Agostino Pierro
Co-Editor in Chief of Pediatric Surgery International
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Preface

Pearls and Tricks in Pediatric Surgery encompass the wide range of complex pediatric surgical issues.

The content presented in this book should be considered as an additional core knowledge not only for the surgeon but also the pediatrician, the gastroenterologist, the neonatologist, the nurse, the surgical trainee, and the medical student.

The scope of this book is not to replace the information in a regular pediatric surgical textbook but to supplement it. Developed from the learner's standpoint, the questions and answers include clinical presentation of the diseases, essentials of pathophysiology, treatments, and possible outcomes.

Pearls are formed inside a shell as a defense mechanism against a potentially threatening irritant. The pearls of knowledge provided in this book should help all caretakers improve the care provided by enhancing their defense mechanisms against complex pediatric surgical issues.

A trick is an effective or quick way of doing something. This book uses the question–answer teaching strategy as an effective vehicle to facilitate learning. It is an old strategy developed by the famous philosopher Socrates who considered the question as the key to all educative activities above the habit-skill level. Its strategy is focused on to achieve the cognitive objectives and bring knowledge to the conscious level.

The authors of the various chapters provide state-of-the-art knowledge based on the current literature, evidence, and personal experience. The quality of the chapters reflects their interest, enthusiasm, and true dedication to learning and teaching the medical school classes and surgical residencies in their institutions. It is an honor, privilege, and a continuing stimulus to be a part of this group of dedicated colleagues. We want to thank them for their excellent job.

We could not have completed this project without the support and understanding of our families with their tireless support and devotion.

Leipzig, Germany
Kansas City, USA
Toronto, Canada

Martin Lacher M.D.
Shawn D. St. Peter M.D.
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Chapter 1

Evaluation of the Pediatric Surgical Patient



Scott S. Short and Michael D. Rollins

Abstract This chapter provides a brief introduction to the nuances and challenges associated with the surgical evaluation of the pediatric patient.

Keywords Surgery · Pediatric · Neonate

1.1 Introduction

It is often stated that children are not small adults. This aphorism simplifies the complexity involved with assessment of the pediatric patient. Not only do children manifest surgical conditions differently than their adult counterparts they have conditions which are specific to periods of development. Further, common conditions can manifest in a variety of ways dependent upon the child's physiologic and psychological development. The "contrast to adult life is greatest in infancy and becomes progressively less until" they have progressed through puberty [1].

1. How are pediatric patients different from adults?

Childhood can be divided into different time periods: neonatal, infant, toddler, child, and adolescent. Each of these developmental periods impart different physiologic and psychosocial features.

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- a. The neonatal period (<30 days of life) is characterized by unique fluid, electrolyte, metabolic, and thermoregulatory requirements. These factors are further influenced by gestational age, birth weight, prenatal factors, and co-morbid conditions. Examples include high resting energy expenditure and glucose requirements, which not only change during the 1st week of life but are directly affected by size and gestation. Clear understanding of these issues as well as unique neonatal conditions (e.g. duodenal atresia, necrotizing enterocolitis) is required by the surgeon.
- b. Infancy (30 days to 1 year of life) is characterized by a period of rapid growth and developmental change. Children often double their birth weight by 6 months of life and see progressive development of motor and social skills. This period is also characterized by unique surgical conditions (e.g. pyloric stenosis).
- c. Toddler (1–3 years) is one of the most challenging periods as communication with the child is difficult and fear of medical personnel often prevents a reliable physical exam. Unique conditions such as intussusception may be seen in this period.
- d. Childhood (4–12 years) reflects continued development towards adulthood. Psychosocial implications of surgery can be quite distressful and many children may experience anxiety and/or regression of developed skills.
- e. Adolescents (>12 years) more commonly have adult type conditions but psychosocial aspects must be considered to develop healthy physician-patient relationships to foster trust, and treatment compliance.

2. What about the parents?

Parents and providers of children are critical in understanding the child's history, presentation, and context of their condition. Young children are non-verbal or often unable to effectively communicate their problems. Older children may not share important information and be resistant to interacting with providers. It is therefore, critical to develop a relationship with the parent not only to understand the clinical problem but to alleviate or address the concerns of the parents themselves. Parental anxieties are known to exacerbate anxiety in the child. Having a sick or injured child is stressful for caregivers and the surgeon must consider their needs in addition to the patient's needs.

3. What do I need to know? What are the important aspects of the history?

In the young child an understanding of the prenatal period is critical. It is important to understand details surrounding the pregnancy (prenatal care, known anomalies, maternal medication use, maternal infections, maternal co-morbid conditions), birth (gestational age, meconium present, rupture of membranes, APGAR scores), and early perinatal course.

Beyond the perinatal period much of the assessment is similar. A complete evaluation of the history of present illness, past medical/surgical history, prior anesthesia history, a family history, social history and physical examination appropriate for the clinical scenario.

Children may present with rare or unusual conditions that may require multi-disciplinary discussion to fully understand the complete picture and to ensure adequate components of the history have been obtained.

Caveats: The family history may be more impactful in some patients and may clue the provider into disease process with known genetic or heritable components. Example: A 2 y.o. girl with a cystic lung lesion. It is important to assess the family for other lesions such as cystic nephroma, multinodular goiter, mesenchymal hamartoma of the liver, etc. to evaluate risk for DICER1 mutations. If positive, one would be concerned that the cystic lung lesion could represent pleuropulmonary blastoma.

4. **How do I examine a child? How is this different?** [2]

a. The assessment:

- i. Child life resources with toys and electronics to distract young children may be helpful in performing an assessment.
- ii. Garnering parental/caregiver support and involvement may not only be necessary but may also allow for a more complete physical examination.
- iii. It is important to try to make the child feel comfortable and safe. This may include examining the child while a parent holds them and/or actively engages them in the assessment. It is also helpful to let the child know what you are going to do next.
- iv. Occasionally, adequate assessment requires an evaluation in the operating room. Examples include: the developmentally delayed child who is too large to safely restrain to adequately evaluate a perianal lesion or a teenage girl who may be too uncomfortable to relax for evaluation of a pelvic straddle injury.

b. The child's size, age, and disease process may affect the exam. Example:

- i. In young infants suspected inguinal hernias can be quite difficult to appreciate on exam. There are many cases where one cannot identify the hernia on exam but the history remains highly suspicious. In these cases of uncertainty, obtaining photos of the hernia can provide confidence before proceeding to surgical repair.

5. **What are requirements/considerations for children undergoing surgical procedures?**

a. Fasting recommendations: [3]

- i. Clear liquids 2 hours
- ii. Breast milk 4 hours
- iii. Infant formula 6 hours
- iv. Full meal 8 hours.

b. If tasked with intubation how do I decide on the appropriate sized endotracheal tube?

- i. Rule of thumb (Child > 2 years): $ETT\ size = (Age + 16) / 4$
- c. Do children have different physiologic considerations important to surgery?
 - i. Yes, Cardiac output varies by age:
 - 1. Neonate: 350 ml/kg/min, infant 150 ml/kg/min, Adult 75 ml/kg/min.
 - ii. Yes, they have increased oxygen consumption and alveolar ventilation
 - 1. Preterm infants may have 3 fold increase in oxygen consumption and children have increased respiratory rates compared to adult counterparts.
 - iii. Yes, they have increased vagal sensitivity
 - 1. More frequent bradycardic events with airway stimulation.
- d. Premature infants can have apneic events. How long do they need to be observed?
 - i. Premature infants (<37 weeks) who are 60 weeks or less post menstrual age require overnight observation
 - ii. Term infants <44 weeks post menstrual age require observation in a monitored bed at least 4 hours
 - iii. Term infants >44 weeks with no history of neonatal apnea can be discharged after meeting discharge criteria.

6. Is routine laboratory evaluation required?

The majority of healthy children undergoing routine outpatient procedures do not require pre-operative laboratory evaluation. For children with medical co-morbid conditions, surgical judgement should be used to decide relevant laboratory values to guide management and limit procedures on children. Examples include obtaining metabolic panels to evaluate the chloride, bicarbonate, and potassium levels on children prior to pyloromyotomy for pyloric stenosis, evaluation of hemoglobin S component on children with known sickle cell disease, and obtaining pregnancy tests in adolescent girls prior to surgery.

7. The parents are worried about anesthesia risk and surgery. How safe is it?

Overall mortality rates have been estimated at less than 1 in 45,000 and many of these occur in children with ASA scores of three or higher [4].

8. How do I effectively communicate surgical planning with a difficult family?

- a. Find a private area for discussion and where distractions can be limited
- b. Discuss the indications, risks, and benefits in a way the family can understand.
 - i. **Don't be rushed**

- ii. It may be helpful to set aside extended time for challenging families (e.g. last clinic patient of the day).
- c. Involve partners and colleagues in the decision making process. “We discussed this as a group and this is what we think will best help your child” or “I spoke to several of my colleagues at other centers and they support the recommendation”.
- d. Utilization of pictures, diagrams, or slides may be helpful in getting the family to understand the proposed procedure. An example may be a PowerPoint slide to illustrate the Nuss procedure for pectus excavatum.
- e. Ask the family (and **patient**) what concerns they have and what they think may help.
- f. Offer a second visit to review ongoing concerns or questions.
- g. Offer to facilitate a 2nd opinion from another provider.

References

1. Bolling RW. Surgery of childhood. New York: Appleton; 1928. p. 1928.
2. Lerwick JL. Psychosocial implications of pediatric surgical hospitalization. *Semin Pediatr Surg.* 2013;22(3):129–33.
3. Practice guidelines for preoperative fasting and the use of pharmacologic agents to reduce the risk of pulmonary aspiration: application to healthy patients undergoing elective procedures: an updated report by the American Society of Anesthesiologists task force on preoperative fasting and the use of pharmacologic agents to reduce the risk of pulmonary aspiration. *Anesthesiology.* 2017;126(3):376–93.
4. Flick RP, Sprung J, Harrison TE, Gleich SJ, Schroeder DR, Hanson AC, et al. Perioperative cardiac arrests in children between 1988 and 2005 at a tertiary referral center: a study of 92,881 patients. *Anesthesiology.* 2007;106(2):226–37; quiz 413–4.

Chapter 2

Nutrition, Fluids and Electrolytes for the Pediatric Surgical Patient



Simon Eaton

Abstract Nutritional care of surgical infants and children is of major importance. This is for several reasons: (i) body stores are often smaller and more precarious; (ii) infants and children not only require energy for maintenance, but also for growth; and (iii) as in adults, recovery from surgery is faster in those patients who are adequately nourished. Surgery for congenital and acquired gastrointestinal anomalies often results in a period during which enteral feeds are not tolerated or absorbed, and provision of parenteral nutrition is often necessary in these patients. However, parenteral nutrition is associated with complications, and should be given only until infants and children are able to tolerate enteral feeds. Fluid and electrolyte balance of surgical infants and children is similarly crucial, because growth and physiological changes in body composition may mask dehydration, edema and electrolyte imbalances exacerbated by losses from the gastrointestinal tract.

Keywords Growth · Nutrition · Parenteral nutrition · Sodium · Stoma

1. How do energy stores in the body alter with age?

Energy stores are only adequate for ~2 days at 24–25 weeks gestation, increase to ~20 days at term as glycogen and fat stores increase and are in excess of 50 days in the adult, hence the urgent need for adequate caloric intake in preterm infants after birth. Full-term neonates have higher content of endogenous fat (approximately 600 g) and therefore can tolerate a few days of undernutrition.

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2. What is the optimum nutritional route for infants?

The optimum nutritional route is oral enteral feeding. However, artificial enteral feeding or parenteral nutrition (PN) may be required if adequate oral feeds cannot be tolerated. The basic principle underlying choice of feeding routes is that the most physiological route that is safely possible should be used: oral preferred over tube feeding, gastric feeds are preferred over jejunal feeds, enteral feeds are preferred over parenteral feeds etc.

3. How should the nutrition of surgical infants and children be monitored?

Effectiveness of nutrition should be assessed. Growth of all paediatric surgical patients, especially those receiving artificial nutritional support, should be monitored longitudinally using appropriate charts. Although measurement of weight, height/length, and head circumference is important, it is essential that these are monitored serially, and plotted on centile charts., which are often available on a national basis, or if not, are available from the World Health Organization. It is especially important to also consider hydration, as over- or under- hydration can be an important contributor to weight change.

4. Why can't premature infants be fed orally?

The swallowing reflex is not fully developed in premature infants so they should be fed by naso- or oro- gastric tubes until the swallowing reflex is developed and it is safe to give oral feeds.

5. Why are gastric enteral feeds preferred over jejunal feeds?

Gastric feeding is preferable to intestinal feeding because it allows for a more natural and complete digestive process i.e. allows action of salivary and gastric enzymes and the antibacterial action of stomach acid, in addition to the use of the stomach as a reservoir. Gastric feeding is associated with a larger osmotic and volume tolerance and a lower frequency of diarrhea and dumping syndrome. Thus, transpyloric feeds are usually restricted to infants or children who are either unable to tolerate naso- or oro- gastric feeds, at increased risk of aspiration; or who have anatomical contra-indications to gastric feeds.

6. Why is long-term nasogastric or orogastric feeding not recommended?

In infants requiring gastric tube feeding for extended periods (e.g. more than 6–8 weeks) it is advisable to insert a gastrostomy, to decrease the negative oral stimulation of repeated insertion of nasal or oral tubes.

7. When should cow's milk protein allergy be considered?

Cow's milk protein allergy can be acute (IgE-mediated) or delayed (non-IgE mediated). Gastrointestinal symptoms are usually present (reflux, colic, constipation etc.), and intolerance in the absence of anatomical reasons may be a manifestation of Cow's milk protein allergy. It can be present even in exclusively breast-fed infants, as bovine antigens may be passed from the mother.

8. What are the advantages of minimal enteral (trophic) feeding?

Minimal feeds may prevent gut mucosal atrophy, increase intestinal blood flow, improve activity of digestive enzymes and thus 'prime' the gut for subsequent higher volume, nutritive feeds. In addition, oral stimulation may prevent later oral aversion.

9. If infants and children are tolerating full feeds, should weight monitoring cease?

Tolerance is not the same as absorption, as infants and children may require a significant period of time for intestinal adaptation to allow complete absorption of administered feeds. Growth monitoring should continue and be checked against centile charts at outpatient follow-up.

10. What might explain poor growth in an infant with a stoma?

Sodium is essential for growth, so that infants with a stoma may have inadequate sodium intake. Low urinary sodium with normal serum sodium suggests active sodium conservation, and sodium supplementation may be appropriate [1].

11. When should parenteral nutrition (PN) be given to a surgical infant or child?

PN is given when enteral feeding is impossible, inadequate, or hazardous, but should be given for the shortest period of time possible and the proportion of nutrition given enterally increased as tolerated. Energy reserves are such that stable term infants can tolerate 3–4 days without enteral feeds, and older children 7–10 days, before starting PN, if it is anticipated that enteral nutrition may be resumed within this time. Premature neonates have smaller energy reserves and the time before introducing PN is much shorter. The most frequent indications in paediatric surgery are intestinal obstruction due to congenital anomalies, although acquired conditions such as post-operative ileus, necrotizing enterocolitis, short-bowel syndrome, gastroenterological indications, and respiratory co-morbidity may require PN for variable lengths of time.

12. Why should PN not be administered peripherally?

Peripheral administration gives significant risk of complications from hyperosmolar glucose, which can cause vascular irritation or damage and thrombosis. PN should be administered via centrally placed catheters (including peripherally inserted central catheters (i.e. PICC lines), surgically placed central catheters or centrally-placed umbilical catheters) dependent on the vascular access already available and the length of time that PN is anticipated to be needed for [2].

13. Which are the components of PN that should be considered as making up the energetic requirements?

The caloric requirements for PN are provided by carbohydrate [3] and lipid [4]. Protein is required for growth and is not used as a source of calories. The ideal

PN regimen therefore, should provide enough amino acids for protein turnover and tissue growth [5], and sufficient calories to minimize protein oxidation for energy.

14. What lipid emulsions should be used in PN of infants and children?

Although pure soybean lipid emulsions can be used short-term, composite lipid emulsions with or without fish oils should be used for PN lasting more than a few days, as this is thought to help prevent cholestasis, one of the major complications of PN [4].

15. Are the energy requirements on PN similar to EN?

No, energy requirements are approximately 10% lower because calorie losses in stool etc. are minimal.

16. Why does weight often drop in the first few days after birth?

This is a normal physiological change in fluid compartments, resulting in diuresis and weight loss of 5–10%.

17. How are hyponatremia and hypernatremia defined?

Hyponatremia is a serum sodium less than 128 mEq/L in the neonate and less than 135 mEq/L in children; hypernatremia is a serum sodium greater than 150 mEq/L.

18. Why are post-operative infants and children at risk of hyponatremia?

Anti-diuretic hormone is secreted for several days in response to operative stress, which can lead to hyponatremia. In addition, gastrointestinal fluid losses also lead to electrolyte losses. Isotonic rather than hypotonic fluids should be administered to decrease risk of hyponatremia, and gastrointestinal electrolyte losses measured and replaced.

19. Which neonatal acquired emergency of term infants is typically accompanied by dehydration and electrolyte disturbances?

Pyloric stenosis typically presents with dehydration together with hyponatremia, hypokalemia, and metabolic alkalosis, so that appropriate resuscitation and correction of electrolyte balance are essential before surgery is performed.

20. How are respiratory and metabolic acidosis/alkalosis differentiated?

In respiratory acidosis/alkalosis, PaCO₂ is >45 mmHg (acidosis) or <35 mmHg (alkalosis) and treatment is via appropriate respiratory support. In metabolic acidosis/alkalosis, bicarbonate <21 mmol/l (acidosis) or >26mmmol/l (alkalosis). In metabolic acidosis it is useful to check the anion gap [=Na+ –(Cl– + HCO₃–), which is normally 12 ± 2 mEq/l] to understand the underlying cause and correct the existing deficits. It is also important, before treatment with sodium bicarbonate bolus, to check the volemic status because of this condition can be due to a tissue hypo perfusion.

21. When should hypotonic fluids be administered?

Hyponatremia at admission, or post-operatively is relatively common in children, so administration of hypotonic fluids should be reserved only for those with a demonstrated hypernatremia >145–150 mEq/L.

References

1. Mansour F, Petersen D, De Coppi P, Eaton S. Effect of sodium deficiency on growth of surgical infants: a retrospective observational study. *Pediatr Surg Int.* 2014;30(12):1279–84.
2. Kolacek S, Puntis JW, Hojsak I, ESPGHAN/ESPEN/ESPR/CSPEN working group on pediatric parenteral nutrition. ESPGHAN/ESPEN/ESPR/CSPEN guidelines on pediatric parenteral nutrition: venous access. *Clin Nutr.* 2018;37(6 Pt B):2379–91.
3. Mesotten D, Joosten K, van Kempen A, Verbruggen S, ESPGHAN/ESPEN/ESPR/CSPEN working group on pediatric parenteral nutrition ESPGHAN/ESPEN/ESPR/CSPEN guidelines on pediatric parenteral nutrition: carbohydrates. *Clin Nutr.* 2018;37(6 Pt B):2337–43.
4. Lapillonne A, Fidler Mis N, Goulet O, van den Akker CHP, Wu J, Koletzko B, et al. ESPGHAN/ESPEN/ESPR/CSPEN guidelines on pediatric parenteral nutrition: lipids. *Clin Nutr.* 2018;37(6 Pt B):2324–36.
5. van Goudoever JB, Carnielli V, Darmaun D, Sainz de Pipaon M, ESPGHAN/ESPEN/ESPR/CSPEN working group on pediatric parenteral nutrition. ESPGHAN/ESPEN/ESPR/CSPEN guidelines on pediatric parenteral nutrition: amino acids. *Clin Nutr.* 2018;37(6 Pt B):2315–23.

Chapter 3

Chest Wall Deformities



Marcelo Martinez-Ferro, Luzía Toselli and Gaston Bellia-Munzon

Abstract Chest wall malformations include excavated deformities or pectus excavatum (PEX) and carinated deformities or pectus carinatum (PC). These deformities can be mixed defects and they may also be a part of a congenital syndrome such as the Currarino Silverman syndrome. The development in the field of chest wall malformations has been steep in the three last decades and both adult and pediatric surgeons have become specialized in the subject pushing even forward the baggage of knowledge. We aim to introduce the reader in the different aspects related to pectus deformities.

Keywords Pectus excavatum · Pectus carinatum · Chest wall deformities · MIPRE · Minimally invasive surgery · Dynamic compression system · Vacuum bell

1. What are the typical chest wall deformities?

The most common are the caved in sternum or pectus excavatum (PEX) and the protruding sternum or pectus carinatum (PC). When these defects coexist in the same patient they are called mixed deformities. Also, deformities may be asymmetric or part of a syndrome such as the Currarino Silverman syndrome, which is

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characterized by a congenital cardiac malformation and pectus arcuatum, a wide, non-articulated, short sternum.

2. What are the demographic characteristics of pectus excavatum (PEX)?

The incidence of PEX has been traditionally described as 1 in 1000, and comprises 80% of all the pectus deformities. However, and probably due to the rise of non-operative approaches to PC, there has been an increase in referrals of PC patients to pectus clinics in the last decades with a shift in the relative incidences in favor of PC resulting in a ratio of 1:1. The sex distribution is predominantly male, with a 4:1 ratio. A family history is common and connective tissue diseases are more frequently associated with pectus deformities.

3. How is PEX classified?

PEX can be classified as typical and atypical. The typical forms can be classified as localized or diffuse, shallow or deep, and as symmetric or asymmetric (Fig. 3.1). The atypical forms include mixed deformities and the Poland syndrome.

4. How is a patient with PEX studied?

The physical examination will provide information regarding the type of PEX and the probability of success with a non-surgical approach. A vacuum bell connected to a vacuumometer can predict how much pressure is necessary to correct the excavation and if the vacuum bell may be effective (Fig. 3.2). We follow our patients with photos taken from 6 predetermined angles at diagnosis and follow-up.

Fig. 3.1 The concept of pectus excavatum (PEX) comprises a wide variety of excavated deformities with typical and atypical presentations. In this figure a deep, localized, symmetric deformity is depicted



Fig. 3.2 A vacuumeter is used to determine objectively the degree of negative pressure necessary to correct the excavated chest wall deformity. This is an indirect way to evaluate the rigidity/elasticity of the deformity



In surgical candidates, a CT scan is performed to quantify the depth of the defect. Physiologic testing may include stress echocardiography as well as a dynamic cardiac magnetic resonance imaging. A history of metal allergy should be inquired and if uncertain, a nickel allergy test has to be performed to determine the patient will tolerate a steel bar. If not, a titanium bar will be needed.

We generally employ a 3D scanning system with virtual reconstruction for diagnosis and follow-up with a visual color-scale that varies according to its depth (Fig. 3.3).

5. What indices are most commonly used to measure the severity of PEX?

The Haller index is the original measure, and it results from the ratio between the lateral distance and the anteroposterior distance between the sternum and the spine, calculated by means of a chest CT scan at the point of maximum sternal depth. This index was not originally validated and it is highly variable depending on sex, symmetry, the shape of the thorax, and the respiration phase in which the study is acquired. Nowadays, the Correction Index has received validation and is more precise to discriminate affected from non-affected subjects.

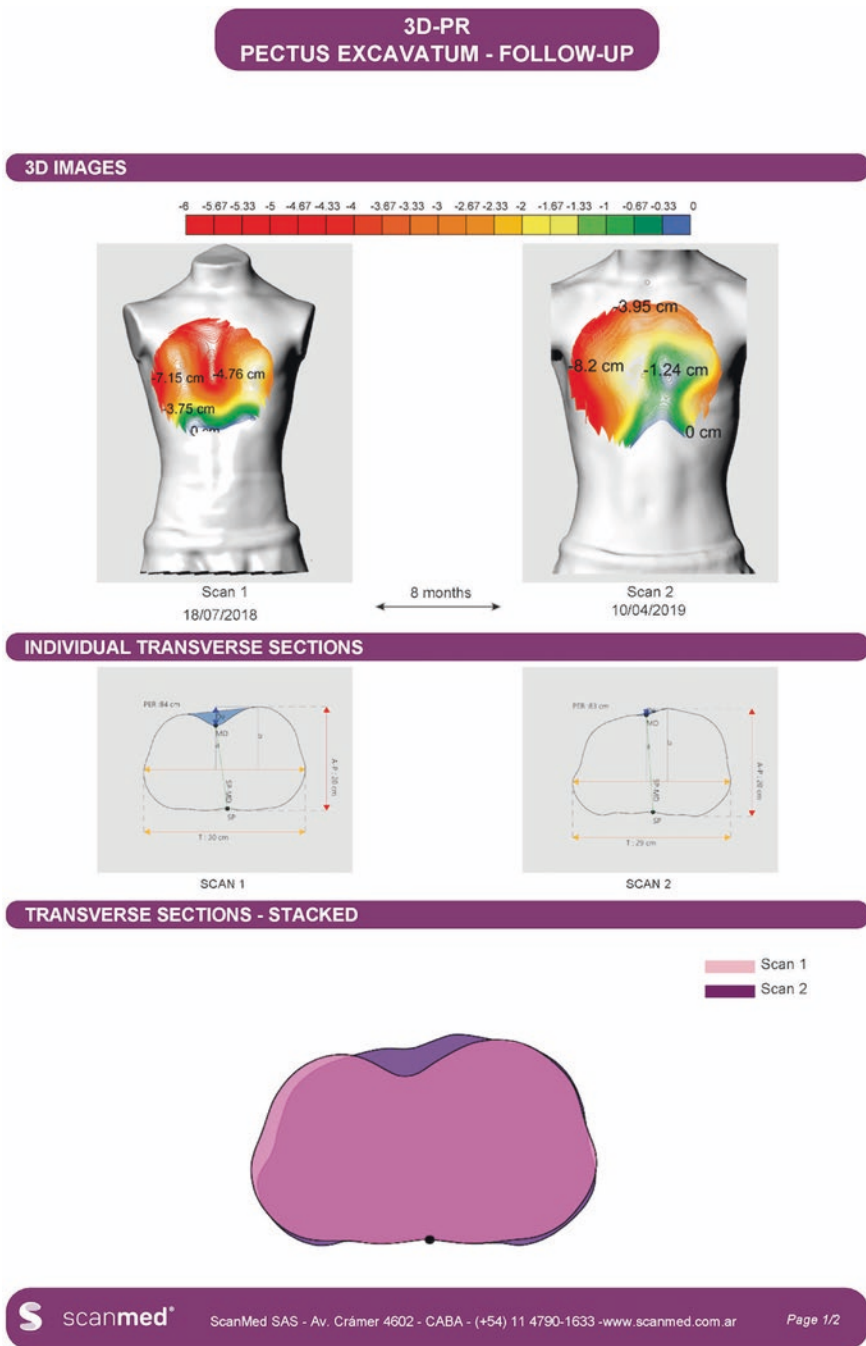


Fig. 3.3 A 3D scanner is used for follow-up. This tool is non-invasive, it does not require radiation, is available at the office and relatively inexpensive when compared to other imaging methods. In this example, this comparative report shows the difference between pre and postoperative transverse section of the chest at the site of maximum depression. Different indexes are determined. In a virtual reconstruction of the anterior chest wall, colours allow an easy interpretation of the geometry of the chest

6. What is the impact of PEX in the cardiopulmonary function?

In echocardiographic studies, functional alterations can be found such as ventricular dysfunction during exercise. Employing dynamic cardiac magnetic resonance, 76% of the patients with PEX have shown some degree of right ventricular compression. Recently, reports have demonstrated a relationship between sternal torsion and cardiac compression as well. Normalization of cardiac function and structure has been reported after PEX repair. More studies are currently underway.

7. What strategies are there for the treatment of PEX?

There are operative and non-operative treatments for patients with PEX. In most cases, chest wall flexibility determines whether surgery will be necessary to solve the deformity. Chest wall flexibility can be determined manually or utilizing a vacuumometer. However, some patients have very dysplastic sternums or rib cages that may require an operation no matter the flexibility they may have.

8. What is the non-operative treatment for PEX?

Since the first report in 2005, the use of a Vacuum Bell has become an option for patients with flexible rib cages [1]. This device is usually appropriate for patients under 11 years old with pectus depth less than 1.5 cm and with good compliance, who wear the device as many hours per day as possible. It is noteworthy that an adjustment period of approximately 6 months of the presternal soft tissue is usually needed to avoid local lesions.

9. In patients with an indication of surgery, what approaches can be used?

Donald Nuss reported the minimally invasive placement of a retrosternal bar in the '90 s. While many variants are described, open resection of costal cartilages is rare today and is generally reserved for extremely asymmetric or mixed deformities.

10. What safety measures should be applied during PEX surgery?

Thoracoscopic guidance is most commonly employed to decrease the possibility of undetected cardiac lacerations when introducing the tunnel dissector. Upward traction on the sternum by vacuum bell or even a crane can be used (Fig. 3.4). A subxiphoid finger can also be used to guide the bar and protect the heart [2].

11. What is the role of cryoanalgesia in pain control of PEX?

Cryoanalgesia has recently become a promising strategy for pain control during and after PEX repair. Thoracic cryoanalgesia consists of the transitory demyelination of the 3rd to the 7th intercostal nerves by the application of a cryoprobe for 2 minutes each at -70 degrees Celsius (Fig. 3.5). Randomized trials have demonstrated that cryoanalgesia is superior to thoracic epidural and patient controlled analgesia in terms of length of stay and requirement of complementary opioids.

12. How long does the bar stay in?

Usually 2.5–3 years.



Fig. 3.4 A crane is our preferred option to lift the sternum prior to retrosternal passage of the dissector. This strategy increases the retrosternal space decreasing the possibility of tears to the pericardium or lacerations to the heart. The sternum is pulled-up via a Lewin surgical clamp exerting traction through small lateral incisions

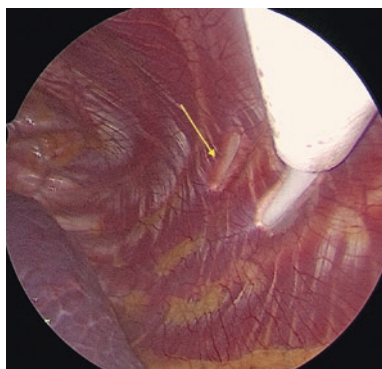


Fig. 3.5 Cryoanalgesia is becoming one of the most interesting strategies for pain control during and after PEX repair. A cryoprobe is introduced to each hemithorax under thoracoscopic guidance. Selective ventilation is performed and the posterior aspect of the intercostal spaces are exposed. The cryoprobe is applied for 2 minutes each space at -70 degrees Celsius. Note the yellow arrow pointing at the previous site of cryoablation

13. What complications may be found during follow-up?

Pneumothorax, pleural effusion, and metal allergy. The more serious complications are bar infection and displacement of the implant. The worst complications are related to cardiac or aortic injury during or after PEX repair.

14. How is Pectus Carinatum (PC) classified?

PC is classified in chondrogladiolar and chondromanubrial types (Fig. 3.6). If the protrusion involves the caudal third of the sternum, it is called chondrogladiolar, the most frequent variant. The chondromanubrial type is a protrusion of the proximal segment of the sternum and frequently comprises a pectus arcuatum, an atypical variant consisting of a wide, short, unsegmented sternum. If it is associated with a cardiac anomaly it is called Currarino Silverman Syndrome. Also, PC may be classified in symmetrical, asymmetrical and mixed.

15. How is a patient with PC studied?

The evaluation of a patient with PC consists of a physical examination with medical photography destined for diagnosis and follow up. CT scans or X-rays are reserved for special cases in which association with skeletal malformations are suspected.

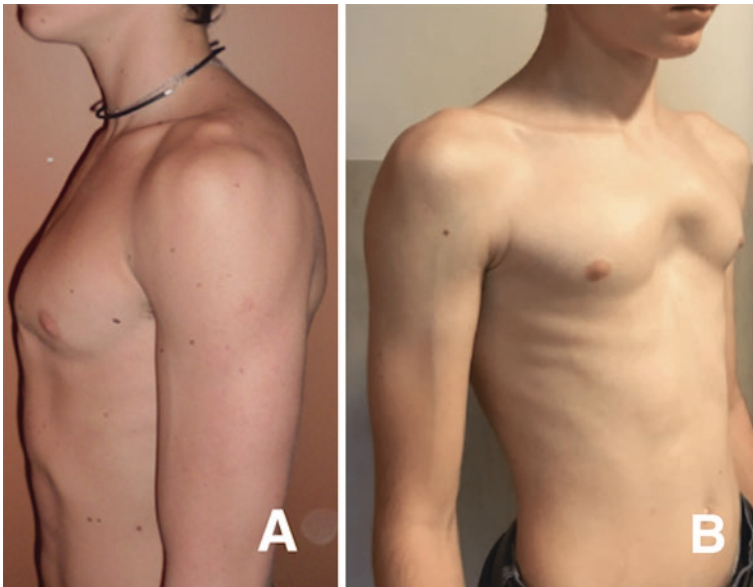


Fig. 3.6 The two most frequent variants of pectus carinatum (PC) are shown. **a** is a teenager with a chondrogladiolar PC in whom the protrusion involves the lower third of the sternum. **b** shows a teenager with a chondromanubrial PC or pectus arcuatum, a very severe form of carinated deformity