MINISTRY OF HEALTH OF UKRAINE
Ukrainian Medical Stomatological Academy

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on meeting the
department of Pediatric Surgery
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METHODICAL INSTRUCTIONS

FOR STUDENTS` SELF-WORK

WHILE PREPARING FOR PRACTICAL LESSONS

Educational discipline	Pediatric Surgery
module №5	Congenital Anomalies in Children
Theme of the lesson	Congenital anomalies responsible for respiratory insufficiency: Malformations of the trachea, bronchi and lungs. Atresia and fistula of the esophagus. Diaphragmatic hernia.
Course	VI
Faculty	foreign students preparation

POLTAVA 2020

1. The topic basis: Relevance of the problem.

The topic "Congenital anomalies responsible for respiratory insufficiency: malformations of the trachea, bronchi and lungs. Atresia and fistula of the esophagus. Diafragmatic hernia. " is very important for future doctors in their professional activity, positively influences the students in their attitude to the future profession, forms professional skills and experience as well as taking as a principle the knowledge of the subject learnt. Actuality of theme is conditioned prevalence of disease, plenty of diagnostic errors, complications and unsatisfactory results of treatment.

2. The aims of the training course:

1. To master classification of malformations of the respiratory system.
2. To recognize the basic clinical displays of malformations which are responsible for respiratory insufficiency.
3. To differentiate malformations of the respiratory system.
4. To interpret the auxiliary methods of research : sounding of gullet, ultrasonic research, radiography (survey, contrasting), bronchograpy, bronchoscopy, computer tomography, magnetic resonance, angiography, and others, laboratory and biochemical analyses, indexes of hemodynamics, immunological.
5. To show examination of child with the malformation of gullet, diaphragmatic hernia and lobar emphysema.
6. To identify the features of motion of congenital malformations of respiratory system.
7. To offer the algorithm of action of doctor and tactic of treatment of patients with congenital malformations of respiratory system.
8. To interpret general principles of treatment of congenital malformations of respiratory system.

3. Basic knowledge, skills, habits necessary for studying the subject (interdisciplinary integration).

Names of previous disciplines	**Obtained skills**
1. Anatomy	Identify and describe the anatomical structure of lungs in children.
2. Faculty pediatrics	Identify and apply additional research methods needed to establish a diagnosis, evaluate the findings.
3. Surgical diseases, topographic anatomy . Operative surgery	Sketch the topography of the chest. Demonstrate the technique of performing pleural puncture. Identify the main priorities of minimally invasive research methods.
4.Propedeutics of childhood diseases	Describe the medical history of sick children with diseases of the pulmonary system, recognition of acute respiratory failure through external examination, palpation, percussion of auscultation.
5.Department of Physiotherapy	The use of physiotherapy

6.Department of Pharmacology	The use of pathogenetic and symptomatic therapy.
7. Radiology	To make an x-ray study, to evaluate the results obtained, to determine the basic radiographic symptoms. Evaluate the data of ultrasound, computed tomography depending on the nature of the pathology

Thereotical questions for the lesson:

1. Embryogenesis of congenital lung cysts.
2. Clinical progress of congenital lung cysts.
3. Clinical manifestations of cystic lung disease
4. Treatment of congenital lung cysts.
5. The etiology of congenital lung emphysema.
6. Clinical manifestations and forms of congenital lung emphysema.
7. Treatment of congenital lung emphysema.
8. Esophageal atresia and ' VACTERLì-'association.
9. Forms of esophageal atresia and its clinical manifestations.
10. Diagnostic of esophageal atresia.
11. Treatment of esophageal atresia.
12. Classification of diaphragmatic hernias.
13. Clinical manifestations of diaphragmatic hernias. "asphyctic infringement".
14. Diagnostic and differential diagnostic of diaphragmal hernias.
15. Operative treatment of diaphragmatic hernias

4. Maintenance of the subject:

PULMONARY APLASIA AND HYPOPLASIA

Pulmonary aplasia results from the interrupted development of the normal bronchial tree with either absence of. or reduction in, the number of normal alveoli. Pulmonary hypoplasia refers to the reduction in size of an entire lung and its individual components. Although these lesions generally arise for different reasons, the former as a primary defect in organogenesis and the latter secondary to extrinsic compression from an intrathoracic mass lesion, they are physiologically similar and are therefore presented jointly here. Although primary pulmonary hypoplasia does occur spontaneously, this problem is more often the result of lesions such as congenital diaphragmatic hernia or CCAM, which limit alveolar development in utero. These children present with newborn pulmonary hypertension, persistent fetal circulation, and respiratory failure.

Several forms of congenital thoracic dystrophy produce acute or chronic asphyxiation related to pulmonary hypoplasia. All are rare, but Jejune thoracic dystrophy is the least rare. The physiologic problem, pulmonary hypoplasia, results from in utero restriction of lung development by an abnormal chest wall. Most affected infants have many other problems and do not survive. The only circumstance in which surgical intervention appears rational is in potentially nonlethal forms of the disease. In this circumstance, procedures designed to enlarge the thorax have been attempted. Median sternotomy and several individualized forms of thoracoplasty have been described. Insufficient data are available for meaningful clinical analysis of these approaches.

CONGENITAL LOBAR EMPHYSEMA

Congenital lobar emphysema, or congenital lobar overinflation. refers to the abnormal postnatal collection of air within a lobe of the lung that is otherwise anatomically normal. This

condition is characterized by expiratory air-trapping within the affected lobe, resulting in lobar parenchymal distention. Compression of adjacent normal lung and mediastinal structures is expected, and physiologic impairment of gas exchange is common. The process is classically the result of developmental deficiency of the cartilage that supports the bronchus to the involved lobe, resulting in focal bronchial collapse and obstruction to expiratory air flow. This specific defect, however, is demonstrable in only one-third to two-thirds of surgically resected emphysematous lobes. The remainder of these infants and children have a variety of partially obstructing bronchial lesions. Some are endobronchial in origin and potentially reversible (e.g.. viscid secretions, mucous plugs, or granulation tissue). Some are the result of extrinsic compression of the bronchus with partial obstruction. Causes include mediastinal lymphadenopathy; adjacent vascular structures, such as an aberrant or enlarged pulmonary artery or ductus arteriosus; mediastinal cysts or tumors that are bronchogenic in origin; or other congenital or acquired mediastinal lesions with an intimate hilar relation. For these reasons, preresection bronchoscope evaluation is recommended with the expectation that reversible bronchial obstruction be corrected before sacrificing a lung lobe that is otherwise normal.

In addition, developmental abnormalities of the alveoli or the terminal airways may be associated with the clinical findings of congenital lobar emphysema. Of note in this regard is the finding of *polyalveolar morphology*, a descriptive histologic term that refers to a substantial and abnormal increase in the number of alveoli present. In this circumstance, postnatal air-trapping occurs within these many alveoli.

Congenital lobar emphysema is a rare lesion, with a 2:1 or 3:1 male predominance. It is most common in the white population. Unilobar involvement is the rule, with affected sites distributed in the following manner: left upper lobe, 40% to 50%; right middle lobe. 30% to 40%; right upper lobe, 20%; lower lobes, 1%; and multiple sites the remainder. Congenital lobar emphysema is associated with congenital heart disease or abnormalities of the great vessels in about 15% of infants. Indeed, extrinsic bronchial compression from vascular structures appears to be a common etiologic problem in this circumstance. For this reason, screening echocardiography is appropriate in all infants with congenital lobar emphysema.

Affected infants usually do not have symptoms at birth. With the onset of extrauterine life and spontaneous respiration, air-trapping and progressive lobar distention develop. Initial clinical symptoms are generally tachypnea and dyspnea, followed by cyanosis if oxygenation is sufficiently impaired. A cough or wheezing may also be present, but this is of little specificity. About one-half of affected infants develop symptoms in the first few days of life; the remainder develop symptoms within the first 6 months. Older infants and children may have few or no symptoms. Infants may have rapidly progressive respiratory failure, with up to 10% to 15% of patients requiring emergency thoracotomy. Generally, the clinical progression is slower, and some patients remain without symptoms. The clinical presentation may be one of progressive respiratory distress; therefore, an affected infant may become increasingly agitated, anxious, and tachypneic. These normal responses to hypoxemia exacerbate the air-trapping phenomenon as the peak inspiratory and expiratory pressures escalate. In particular, focal bronchial collapse occurs with excessive expiratory effort; as this develops, the lobar emphysema worsens, and further compromise in gas exchange results. Likewise, positive-pressure ventilation can induce acute lobar distention with potentially catastrophic respiratory decompensation or mediastinal displacement. The physiologic derangements may be indistinguishable from those of tension pneumothorax. This is an important consideration during endoscopic evaluation of the endobronchial tree. Particularly in infants with preoperative symptoms, the surgeon must be prepared to decompress the thorax by emergent thoracotomy and then to proceed with definitive lobectomy.

Congenital lobar emphysema is typically found in term infants, but acquired emphysematous disease in preterm infants is common. The cause of this latter problem is multifactorial, including barotrauma from positive-pressure ventilation, oxygen toxicity, and lung immaturity. It is often seen in conjunction with, and as a complication of.

bronchopulmonary dysplasia. Unlike congenital lobar emphysema, multiple areas of focal hyperinflation and interstitial emphysema are often present. Unilobar right lower lobe involvement is also common, probably as a consequence of endotracheal tube positioning, which selectively ventilates the right mainstem bronchus. These characteristics help differentiate congenital and acquired disease.

Physical findings of congenital lobar emphysema may include an asymmetric thorax, a shift in the apical cardiac impulse to the contralateral side, and focal hyperresonance and diminished breath sounds over the affected lobe. None of these findings, however, has the necessary sensitivity and specificity to demonstrate the precise nature of the problem.

The diagnosis is best established by plain chest radiograph. Typical findings include lobar hyperinflation, contralateral shift of the mediastinum and trachea, compression or even lobar atelectasis of adjacent lung, and flattening of the ipsilateral hemidiaphragm. If these findings are all present, there is no need for additional imaging studies. Differentiating this presentation from tension pneumothorax is essential. The latter is characterized by collapse of the entire affected lung into the hilum. In contrast, although lobar emphysema can be dramatic in its radiographic appearance, adjacent compressed lung can almost always be discerned, most often the lower lobe at the base of the thorax. In addition, the occasional congenital cystic adenomatoid malformation (CCAM) with a single large cystic component can be mistaken for lobar emphysema. Because lobar emphysema rarely involves the lower lobes, this is an important differentiating feature. Nonetheless, the surgical management of these latter two lesions is similar so preoperative differentiation is less important than for tension pneumothorax, for which the treatment is different. As with most mass lesions in the chest, computed tomography (CT) and magnetic resonance (MR) imaging provide excellent anatomic information for infants with congenital lobar emphysema. These procedures are most helpful in elective situations when the diagnosis is in doubt. In addition, ventilationB"b"perfusion scans have been employed to evaluate infants with lobar emphysema, particularly when the areas of involvement are multiple or the disease acquired. In this setting, specific areas of nonfunctional lung can be identified and resected if they appear to compromise adjacent normal lung.

Because the natural history of congenital lobar emphysema is often progressive and includes potentially life-threatening respiratory insufficiency, prompt surgical lobectomy is the treatment of choice for infants and young children. Because the underlying lesion is structural, medical treatment can be considered only a supportive adjunct in patients with symptoms. In patients without symptoms, particularly older children, this approach may be tempered reasonably because the likelihood of sudden decompensation in this circumstance is low. The rationale for routine endoscopic evaluation of the affected bronchus has been noted. The purpose is to identify and eliminate reversible endobronchial obstructions from secretions, mucous plugging, or granulation tissue. Clearly reversible endobronchial problems should be corrected without parenchymal lung resection.

Extrinsic bronchial compression is associated generally with a focal cartilaginous defect of the affected bronchus that is not adequately relieved by simple decompression.

Although congenital lobar emphysema results from a specific anatomic defect, reconstructive procedures, such as bronchoplasty or segmental bronchial resection and anastomosis, are generally inappropriate. The diminutive size of the infant bronchus and the possibility of nonfocal cartilaginous tracheobronchial defects present important technical obstacles to successful local reconstructive procedures. In addition, there is little reason to select this approach because the clinical results of lobectomy are generally excellent for this lesion.

Acquired emphysema is often seen in preterm infants with a multitude of other problems. Treatment is generally medical and supportive, with the natural history being one of slow resolution over a number of months. In the acute phase, selective ventilation of nonemphysematous areas of lung or the use of alternative strategies such as high-frequency oscillatory or jet ventilation can minimize the peak airway pressure, which is directly correlated to the formation of emphysema. These approaches can also help infants with congenital lobar

emphysema if prolonged transport is necessary or there is delay in reaching the operating suite. Lobectomy may be beneficial in occasional selected patients with severe regional emphysematous disease; however, late death may result from associated bronchopulmonary dysplasia in these patients.

As noted, infants and children have an excellent response to lobectomy for congenital lobar emphysema. Even in those who are critically ill and require emergency thoracotomy, the physiologic response is a predictably prompt and dramatic return to normal after resection of the affected lobe. Mortality for this specific lesion is rare in a modern pediatric surgical environment. The general risks of thoracotomy and lung resection include morbidity related to anesthesia, empyema, pneumothorax, infection, bleeding, and bronchopleural fistula. These are not different than for any other neonatal thoracotomy and lobectomy, and are presented in detail in the section that deals with outcomes after lung resection. The cumulative incidence of these types of complications is about 5% to 10% in most modern pediatric surgical practices, although it has been as high as 20% to 40% in recent decades. Long-term pulmonary function is also predictably excellent after lobar resection, and this is discussed separately later. For infants with coexisting congenital heart disease, acquired pulmonary emphysema, or additional medical problems, the outcome is generally dictated by these other conditions.

In follow-up studies by Frenckner and Freyschuss. Actual lung volumesB"b'residual volume, vital capacity, total lung capacity, and forced expiratory volume in 1 second (FEV,)B"B"in patients who had undergone neonatal lobectomy for congenital lobar emphysema were 90% of predicted values, and no long-term functional impairment was reported. Infants who had undergone neonatal lobectomy for congenital lobar emphysema were evaluated as adults by McBride and colleagues in 1980. Ipsilateral and contralateral lung volumes were found to be equal, despite the previous lobectomy. This appeared to be the result of compensatory tissue growth, not simply distention of residual lung parenchyma. In this latter study, perfusion was found to be equally distributed between the operated and nonoperated lungs. These patients demonstrated diminished expiratory flow rates compared with expected values (FEV,, 72% of predicted; maximal midexpiratory flow, 45% of predicted). These findings appear to result from disproportional growth between the conducting and the terminal airways during infancy. This concept does not diminish the excellent clinical prognosis for these infants, and is presented in detail at the end of this chapter.

BRONCHOGENIC CYSTS AND LUNG CYSTS

A developmental cyst arising from the trachea or a bronchus is referred to as a *bronchogenic cyst.* These account for about 20% to 30% of congenital bronchopulmonary-foregut cystic malformations. Potential locations include the cervical or thoracic trachea, the hilar bronchi, or the more distal intraparenchymal bronchi. It has been reported that about 70% of thoracic bronchogenic cysts are located within the lung parenchyma, and the remainder are in the mediastinum, but this distribution varies considerably among different reports. Ectopic bronchogenic cysts, including those in paravertebral, paraesophageal, pericardial, subcarinal, and subcutaneous locations, have been reported.

Bronchogenic cysts are typically unilocular mucus-filled lesions arising from the posterior membranous portion of the airway. They do not usually communicate with the functional tracheobronchial tree. Many anatomic variations, however, have been described. By definition, the cyst has structural elements of the airway, including cartilage, smooth muscle, mucous glands, and respiratory epithelium. Likewise, these lesions have a normal bronchial arterial blood supply. The character of the epithelium depends on the site of origin; ciliated columnar, cuboidal. and squamous epithelium are all found within the tracheobronchial tree, and therefore, within these cysts.

This section also considers cystic lung lesions that result from abnormal development of the more distal airways, alveoli, or pleural or lymphatic tissue. Even collectively, these true lung cysts are rare congenital lesions. They constitute a heterogeneous group of lung parenchymal cystic lesions with histologic features representative of their sites of origin. The spectrum is

varied and can overlap with cysts that are bronchogenic in origin. Differentiation of the tissue of origin for simple lung cysts is principally of pathologic interest because the presentations are similar, and clinical management is generally straightforward with a good outcome. One important exception is when the developmental abnormality is lymphatic in origin. The result then may be pulmonary lymphangiectasis. This is typically characterized by diffuse bilateral pulmonary cystic disease, and the outcome is often lethal because resection is not feasible.

The discussion of bronchogenic and other lung cysts is consolidated because of the overlap in their clinical presentations and the similarity in their embryologic origins. As with other congenital cystic lung lesions, physiologic injury from bronchogenic and lung cysts generally results from either compression of adjacent hollow viscera, such as the airway or esophagus, or inadequate drainage of secretions with secondary infection. Malignancies have also been reported within these lesions, and rhabdomyosarcoma, bronchogenic carcinoma, and adenocarcinoma have been described. In newborns with cysts adjacent to the trachea or proximal airways, respiratory distress or air-trapping with lobar emphysema are important and potentially life-threatening problems. More distal lesions may be asymptomatic or may present with evidence of infection. The latter usually occur in older children because time is necessary for the development of infection. Clinical presentations range from no symptoms to life-threatening respiratory distress, although the latter is rare. Infection and nonspecific respiratory symptoms. such as cough, dyspnea, tachypnea, wheezing, or chest pain, are typical. The usual chest radiographic appearance of a bronchogenic cyst is that of a smooth, roughly spherical, paratracheal. or hilar solid mass without calcification. Displacement of the adjacent airway and distal air-trapping are relatively frequent, even in patients without symptoms. Air-fluid levels suggest communication with the tracheobronchial tree or foregut. and this is a particularly likely finding in the presence of acute infection.

True lung cysts can occur anywhere. They are typically single, unilocular lesions. These can be large or small and difficult to distinguish from a lung abscess or macrocytic CCAM on chest radiograph. Discovery of a bronchogenic or true lung cyst after slow or incomplete radiographic resolution of acute pneumonia has also been well described. As with other thoracic mass lesions. CT and MR imaging provide both diagnostic accuracy and excellent definition of the anatomic relations of these lesions. In the patient with dysphagia and a paraesophageal bronchogenic cyst, a contrast esophagogram may demonstrate extrinsic compression at the site of the lesion. Likewise, endoscopic examination of the tracheobronchial tree or esophagus may show extrinsic compression.

Resection of the cystic abnormality is standard treatment for virtually all bronchogenic and lung cysts, even if asymptomatic. The risk of infection appears to be high, although no prospective data exist. Generally, simple local resection is easily accomplished and definitive. Occasionally, however, limited parenchymal lung resection or even lobectomy may be required. Preoperative treatment of pneumonia is helpful in diminishing perioperative morbidity and in minimizing the magnitude of parenchymal resection. Preservation of adjacent normal parenchyma is an important operative principle. Wedge resection, segmentectomy. and lobectomy have all been reported for individual circumstances. As with many other thoracic lesions, thoracoscopic resection of bronchogenic and lung cysts is feasible for selected patients. It is essential to establish precise anatomic relations preoperatively if a thoracoscopic approach is planned because bronchogenic cysts are often beneath the mediastinal pleura, and therefore, require pleural incision and mediastinal exploration to localize the lesion. Mediastinal exploration is important for infants with lobar emphysema because an occult bronchogenic cyst may be responsible, and relief is occasionally possible without lobar lung resection.

The long-term outcome for infants and children with bronchogenic and true lung cysts is excellent because they generally do not require sacrifice of significant normal lung parenchyma. Likewise, perioperative morbidity is low and mortality rare, particularly for mediastinal lesions without tracheobronchial communication. If lung resection is required, outcome is not different than for patients with other lung lesions, such as lobar emphysema or CCAM and these

outcomes are presented later in detail.

ESOPHAGEAL ATRESIA/TRACHEOESOPHAGEAL FISTULA

EA is a condition in which the proximal and distal portions of the esophagus do not communicate. The upper segment of the esophagus is a dilated blind-ending pouch with a hypertrophied muscular wall. This pouch typically extends to the level of the second to fourth thoracic vertebra. In contrast, the distal esophageal portion is an atretic pouch with a small diameter and a thin muscular wall; it extends a variable distance above the diaphragm.

TEF is an abnormal communication between the trachea and esophagus. When associated with EA, the fistula most commonly occurs between the distal esophageal segment and the trachea. The distal esophageal segment communicates with the trachea just above the carina. An H-type TEF represents a TEF without EA. It can occur at any level from the cricoid cartilage to the carina, although it usually courses obliquely (with the tracheal end proximal) at or above the level of the second thoracic vertebra.

Background, types of EA and TEF

Five types of EA and TEF have been described. The most common abnormality is EA with a distal TEF (84%). Isolated atresia with no fistula is the next most common finding (8%), followed by TEF with no atresia (so-called H type) (4%). EA with proximal and distal fistulas (3%) and EA with a proximal fistula (1%) are less common.

Diagram depicting the five variations of oesophageal atresia.

(a) Atresia – no fistula (5–10%).

(b) Oesophageal atresia with high fistula only (1%).

(c) Oesophageal atresia with low fistula only (80–90%).

(d) Oesophageal fistula with low and high fistula (2–3%).

(e) H-fistula with no atresia (5–8%).

Etiologies and factors. The etiologies of these anomalies are still largely unknown, but many theories concerning their origins have been proposed. The trachea and esophagus are foregut derivatives. Lateral mesodermal ridges form in the proximal esophagus during the fourth gestational week, and the fusion of these grooves in the midline separates the esophagus from the trachea around the 26th day of gestation.

Notochord abnormalities, desynchronous esophageal mesenchymal and epithelial growth rates, neural crest cell involvement, and incomplete tracheoesophageal separation resulting from a lack of apoptosis are mentioned in some of the theories that have been proposed for EA embryogenesis. Similarly, incomplete tracheoesophageal septation, lateral ridge fusion failure, and tracheal and esophageal proximity have been suggested in explanations of the origin of TEF. In addition, vascular insufficiencies; genetic factors; vitamin deficiencies; drug and alcohol exposures; and viral, chemical, and physical external events may contribute to the development of EA and/or TEF.

Pathophysiology. Because of the discontinuous esophagus, infants with EA cannot clear their secretions. This defect leads to persistent drooling and aspiration or regurgitation of food after attempted feedings. TEF causes additional complications because of the tracheoesophageal communication. When infants with this anomaly strain, cough, or cry, air enters the stomach through the fistula. As a result, the stomach and small intestine become dilated, elevating the diaphragm and making respiration more difficult. The reflux of food and gastric secretions may also occur up the esophagus and through the fistula into the tracheobronchial tree; this reflux can contribute to pneumonia and atelectasis. Therefore, pneumonia and respiratory distress are common complications.

Abnormal esophageal motility has been observed in children with EA and/or TEF. Controversy often exists as to whether the abnormality was inherently present in the child's esophagus or if the dysfunction was a result of the surgical treatment. Manometric studies have shown that the motility disorder is present before surgical treatment. Animal studies have also shown that esophageal transection followed by repair does not precipitate disturbances in motility.

Discoordinated peristalsis has been reported from the level of the fistula to the stomach in patients with isolated TEF.

Frequency. The international occurrence varies, with estimates ranging from 0.4 to 3.6 cases per 10,000 live births in different regions of the world.

*Mortality/Morbidity***.** Despite an increased number of patients with severe anomalies, survival rates as high as 95% have been reported. In uncomplicated cases, survival rates approach 100%.

Clinical signs. The first clinical sign of an infant with EA is maternal polyhydramnios resulting from the infant's inability to swallow and absorb amniotic fluid through the gut. Polyhydramnios is seen in infants with many diagnoses; only 1 in 12 infants with polyhydramnios have EA. Polyhydramnios is seen in 95% of infants with EA and no fistula and in 35% of patients who have EA with a distal fistula. Increased pressure due to the amniotic fluid accumulation results in a greater number of premature births and neonates with low birth. One third of infants with EA weigh less than 2250 g.

Most infants with EA become symptomatic within the first few hours of life, unlike children with an isolated fistula, who have more subtle symptoms that may not be recognized initially. Excess salivation and fine, frothy bubbles in the mouth and sometimes nose result from an inability to swallow. Any attempts at feeding result in choking, coughing, cyanotic episodes, and food regurgitation. The presence of a fistula increases respiratory complications due to aspiration of food and secretions in the trachea and lungs. Pneumonitis and atelectasis develop quickly in the affected neonate, and rattles heard during respirations are common. Fistulas also allow air to enter the stomach and intestines, leading to abdominal distension. With atresia alone, the abdomen appears scaphoid.

Many anomalies are associated with EA, and 50-70% of children with EA have some other defect.

Diagnosis. Once the EA is considered, appropriate diagnostic procedures are necessary. The simplest and quickest diagnostic procedure is the passage of a 10 to 12 French oral tube into the esophagus. If an obstruction encountered (usually in 9 to 13 cm), EA is likely. If the tube passes belong this point, atresia is unlikely. In either case, a radiograph of the chest must be obtained to confirm the position of the tube.

Ultrasound. Although ultrasonography has no role in the routine postnatal evaluation of EA and/or TEF, prenatal sonography is a valuable screening tool for EA and/or TEF. The diagnostic accuracy is increased if an anechoic area is present in the middle of the fetal neck; this sign differentiates EA from diseases with possible swallowing impairments.

Plain radiographs provide much information, including findings for EA confirmation and depiction of the side of the aortic arch side, presence of any vertebral or other associated anomalies, and others. Barium studies performed after the surgical placement of a gastrostomy may be used to evaluate the gap length and associated GI abnormalities such as duodenal atresia or malrotation. However, radiographs may not always demonstrate the presence of a fistula.

Findings on posteroanterior and lateral chest images confirm a diagnosis of EA by displaying a coiled nasogastric tube (placed for determination of EA) in the proximal esophageal pouch of a child with EA.

Any vertebral anomalies may be visualized, and some cardiac anomalies may be suggested. Aspiration pneumonia, especially in the right upper lobe, and patchy atelectasis are frequently present.

Aside from these general findings, the radiographic observations in children with EA and/or TEF vary depending on the type of anomaly present.

H-type of tracheooesophageal atresia.	Oesophageal atresia. Total absence of gas in the bowel.

The anatomy should be clarified ***bronchoscopically*** at the time of the first anesthetic.

Treatment.

Preoperative management.

- Confirming the diagnosis and type of anomaly;
- Evaluating the pulmonary status, treating existing pulmonary problems, and preventing further tracheal contamination;
- Searching for, and treating other major associated problems.

To prevent further aspiration, the pharynx is frequently suctioned. The patient is cared far in a head-elevated position, with a catheter insert into the upper esophageal pouch and connected to continuous sump suction.

All patients are given antibiotic.

If there is clinical or radiological evidence of significant atelectasis or pneumonia, a decompression Stamm gastrostomy is performed using general anesthesia. Patients usually respond well within 24 to 72 hours, at which time the anomaly is repaired.

Gastrostomy decompression, upper pouch suction, and central venous nutritional support allow for the stabilization of the patient, the search for other disease, and the treatment of both medical or surgical problems.

Operative management.

Surgical repair is performed under general anaesthesia with endotracheal intubation. Trachea and main bronchi are briefly inspected, and the fistula to the oesophagus is localized, which is usually approximately 5–7 mm above the carina. Exceptionally, it may be found at the carina or even in the right main bronchus, indicating a short lower segment, and most likely with a long oesophageal gap. The next step is to look for an upper fistula. The dorsal – membranous – region of the tracheal wall is inspected carefully up to the cricoid cartilage. Small upper fistulas are easily missed. To avoid this pitfall, irregularities of the dorsal wall are gently probed with the tip of a 3F ureteric catheter passed through the bronchoscope. If a fistula is present, the ureteric catheter will glide into it.

The standard approach for repair of an oesophageal atresia is a right latero-dorsal thoracotomy. If a right aortic arch is diagnosed pre-operatively, a left-sided thoracotomy is recommended.However, if an unsuspected right descending aorta is encountered during surgery, the procedure can be continued in most cases, establishing the anastomosis on the right of the aortic arch..

Postoperative care.

Antibiotics are continued for several days.

The chest tube is left in place for 10 days.

In the large, healthy baby, the endotracheal tube can be safety removed soon after the procedure. In the small, ill infant, ventilatory support is usually required for several days. Small amounts of clear liquid oral feeding are started when the infant can handle the saliva.

When the patient is taking adequate oral feedings, swallowing without difficultly, and gaining weight, the parents are instructed in home care, and the infant is discharged. A gastrostomy tube, if present, is usually removed before the infant goes home.

Complications. The most common complications include anastomotic leak, recurrent fistula, stricture, and gastroesophageal reflux (GER).

CONGENITAL DIAPHRAGMATIC HERNIA

Congenital diaphragmatic hernia (CDH) is an abnormal opening in the diaphragm that allows part of the abdominal organs to migrate into the chest cavity, occurring before birth.

Congenital diaphragmatic hernia is seen in 1/2200 to 1/5000 live births with the vast majority (80 to 90%) occurring on the left side. There is a 2% recurrence rate in first degree relatives of a patient with the disease.

Diaphragmatic hernia is usually a sporadic abnormality. However, in about 50% of affected fetuses there are associated chromosomal abnormalities (mainly trisomy 18, trisomy 13 and Pallister–Killian syndrome – mosaicism for tetrasomy 12p), other defects (mainly craniospinal defects, including spina bifida, hydrocephaly and the otherwise rare iniencephaly, and cardiac abnormalities) and genetic syndromes (such as Marfan syndrome).

Anatomy of the diaphragm. The diaphragm is the fibromuscular sheet that that separates the thoracic and abdominal cavities; it is the principal muscle of inspiration. The fibrosus portion of the diaphragm, the central tendon, accounts for about 35 % of its total surface. The muscular portion arises from whole of the internal circumference of the thorax, being attached:

1. In front, by fleshy fibres to the xiphoid process,
2. To the internal surface of the 6 to 7 inferior ribs,
3. Behind, to two aponeurotic arches, named the ligamentum arcuatum externum and internum,
4. By the crura, to the to the lumbar vertedrae. The right and left crus arise from the anterior surface of the bodies and intervertebral substances of 4 upper lumbar vertebrae, on each side of the aorta.

In 80 % of bodies, there is the gap between the muscles arising from the ligamentum arcuatum externum and those of the costal origin as they traverse to the central tendon, which is called the lumbo-costal triangle. Bochdalec postulated that congenital posterolateral diaphragmatic hernia was caused by a weakness in the area of the lumbo-costal triangle.

Normally, there are 3 significant openings in the diaphragm:

1. The vena cava transverses the central tendon to the right of the midline;
2. The esophageal hiatus is just to the left of the midline and slightly posterior to the plane of the vena cava;
3. The aorta lies on the verterbral bodies.

Herniation of the viscera through the opening of the aorta or vena cava has not been described. However, herniation of the stomach through the esophageal hiatus is common and incidence increases with aging.

Five defects might develop in the diaphragm to create intraabdominal viscera herniation.

1. The esophageal hiatus is the most frequent area, whrein the stomach pralapses into the mediastinum.
2. A congenital posterolateral defect occurs from maldevelopment of the diaphragm. The Bochdalek CDH accounts for approximately 70% of cases and occurs in a posteriolateral region of the diaphragm.
3. Anomalous attachment of the diaphragm to the sternum and adjacent ribs results in a foramen, which allows the bowel to extend into the anterior mediastinum.
4. The assotiation of an epigastric omphalocele and a retrosternal defect in the diaphragm and pericardium (pentalogy of Cantrell) results in herniation within the pericardium.
5. Attenuation of the tendinous or muscular portion of the diaphragm produces eventration.

Simile to eventration, paralysis of the muscles of the diaphragm either from trauma to the phrenic nerve or a congenital defect in the anterior horn cells of the cervical spine cord (C – 3, 4) as in Werdnig-Hoffmann disease, results in herniation of the intraabdominal contents into the thoracis cavity.

Embriogenesis of posterolateral diaphragmatic hernia and pulmonary hypoplasia. The septum transversum grows posteriorly to meet the dorsal mesentery of the foregut, forming the central portion of the diaphragm during the fourth to eighth weeks of fetal life. The lateral folds of peritoneum and pleura develop simultaneously, completing the separation between the thorax and the abdomen. Disruption of this process results in a posterolateral defect in the diaphragm (foramen of Bochdalek). This closure of the pleuroperitoneal canal is completed on the right side before the left, which may explain why 90 % of the diaphragmatic defects present on the left side.

As the diaphragm is developing, the midgut undergoes elongation and development outside the coelom. The midgut normally returns to the abdominal cavity and undergoes rotation and fixation at about the tenth week of fetal life. If closure of the diaphragm is incomplete, the intestine herniates into the chest as it returns to the coelom, inhibiting development of the lung and preventing the normal process of intestinal rotation and fixation. The spleen, stomach, and left lobe of the liver as well as the bulk of the intestine often reside within the thoracic cavity.

Herniation of the abdominal viscera into the thoracic cavity seriously impairs development of the ipsilateral, and to some extent the contralateral, lung. This hypoplasia of the pulmonary parenchyma accounts for the major physiological problems encountered in these infants.

The degree of hypoplasia of the pulmonary vessels correlates with the adequacy of ventilation. If the pulmonary parenchyma is inadequate, mechanical ventilation will not prevent severe hypoxia, hypercarbia, and acidosis in these infants. Occasionally, adequate ventilation can be achieved initially, indicating that enough pulmonary parenchyma is present to sustain life. Later, arterial blood gas balance deteriorates, with the development of pulmonary hypertension and right to left shunting through the patent ductus arteriosus (persistent fetal circulation). In this situation pulmonary circulation with progressive vasoconstriction and pulmonary hypertension decreases perfusion of the lung and results in clinical deterioration. Vasodilating agents do not alter this pattern of persistent fetal circulation. They dilate the systemic as well as the pulmonary circulation, causing systemic hypotension; this creates a need for additional fluid administration, which in turn results in further pulmonary oedema and deterioration.

After birth air entering the gastrointestinal tract distends the herniated bowel, shifting the mediastinum toward the contralateral side and compressing the contralateral lung. This process can be largely prevented by sump catheter decompression of the stomach.

Clinical manifestations. Most infants develop respiratory symptoms in the first 24 h after birth. Although a spectrum of respiratory distress exists, many children immediately become severely cyanotic. Physical examination discloses a scaphoid abdomen, displacement of the cardiac apex away from the side with the defect, and decreased or absent breath sounds on the affected side. Rarely, bowel sounds can be heard in the chest. The occasional infants who become symptomatic weeks to months after birth respond well to surgical treatment.

Diagnosis. Prenatally, the diaphragm is imaged by ultrasonography as an echo-free space between the thorax and abdomen. Diaphragmatic hernia can be diagnosed by the ultrasonographic demonstration of stomach and intestines (90% of the cases) or liver (50%) in the thorax and the associated mediastinal shift to the opposite side. Herniated abdominal contents, associated with a left-sided diaphragmatic hernia, are easy to demonstrate because the echo-free fluid-filled stomach and small bowel contrast dramatically with the more echogenic fetal lung. In contrast, a right-sided hernia is more difficult to identify because the echogenicity of the fetal liver is similar to that of the lung, and visualization of the gall bladder in the right side of the fetal chest may be the only way of making the diagnosis.

Polyhydramnios (usually after 25 weeks) is found in about 75% of cases and this may be the consequence of impaired fetal swallowing due to compression of the esophagus by the herniated abdominal organs.

After birth, a chest radiograph should be obtained, preferably after an orogastric tube has been passed into the stomach. If the radiograph is taken before air enters the bowel, the affected chest is radiopaque, but the trachea and heart are shifted to the contrlateral side, and the aerated lung is diminished. An upright thoracic radiograph shows multiple loops of intestine in the thoracic cavity and no diaphragmatic outline.

Differential diagnoses include cystic adenomatoid malformation, eventration of the diaphragm, pneumatoceles from staphylococcal pneumonia, and pulmonary agenesis or hypoplasia. The radiographic appearances of all of these entities include presence of the diaphragm and a normal intra-abdominal bowel gas pattern.

Treatment.

Preoperative care. Initial resuscitation is mandatory if these infants are to survive. Intubation for respiratory distress is often required before a radiograph is available. A nasogastric tube should be inserted and placed on suction to prevent further distention of the intestine. Because the hypoplastic lungs are susceptible to barotrauma, ventilation with high pressure must be avoided. A tension pneumothorax on the contralateral side often proves fatal. Rapid ventilation with short inspiratory times, low pressure, and 100 % oxygen is most effective. Alkalosis, hypocarbia, and oxygenation all decrease pulmonary artery pressures and the right to left shunt seen in persistent

fetal circulation. Sedation seems to benefit those with very reactive pulmonary vasculature, but it does not help those with inadequate pulmonary tissue, who develop early pulmonary hypertension and acidosis. The infant must be kept warm during transport to avoid peripheral vasoconstriction and acidosis: the latter can elevate pulmonary artery pressure.

Although surgical repair was traditionally performed urgently, recent studies have shown that ventilatory compliance decreases appreciably in infants after surgery, producing a decline in the arterial gases. The current practice is to stabilize these infants with ventilation, sedation, and intestinal decompression, deferring repair of the defect until 36 to 72 h after birth. Delay may avoid the development of pulmonary hypertension and persistent fetal circulation due to surgical stress.

Some infants are placed on ECMO (extracorporeal membrane oxygenation) which is a heart/lung bypass machine which gives the lungs a chance to recover and expand after surgery.

Surgical repair.

General anaesthesia with muscle relaxation is used. The baby is positioned supine on a warm blanket. The most commonly preferred approach is abdominal. This offers good exposure, easy reduction of the abdominal viscera and recognition and correction of associated gastrointestinal anomalies. A subcostal transverse muscle cutting incision is made on the side of the hernia.

The contents of the hernia are gently reduced in the abdomen. On the right side, the small intestine and colon are first reduced and the liver is withdrawn last. After the hernia is reduced, an attempt is made to visualize the ipsilateral lung. This is usually done by retracting the anterior rim of the diaphragm. Often, a hypoplastic lung can be observed at the apex. A hernial sac, composed of pleura and peritoneum, is present in about 20% of patients. The sac, if present, is excised to avoid leaving a loculated spaceoccupying lesion in the chest.

Most diaphragmatic defects can be sutured by direct sutures of the edges of the defect. Usually the anterior rim of the diaphragm is quite evident. However, the posterior rim may not be immediately apparent and may require dissection for delineation. The posterior rim of the diaphragm is mobilized by incising the overlying peritoneum.

The defect is closed by interrupted non-absorbable sutures. Occasionally, the posterior rim is absent altogether, in which case the anterior rim of the diaphragm is sutured to the lower ribs with either periostial or pericostal sutures.

If the defect is large, it may not be possible to repair it by direct suture. Various techniques have been described and include the use of prerenal fascia, rib structures, the latissimus dorsi muscle, rotational muscle flaps from the thoraco-abdominal wall and prosthetic patches. The operations involving muscle flaps are too long and complex for critically ill patients and can lead to unsightly chest deformities. Prosthetic materials, including Marlex mesh, reinforced silicone elastomer, preserved pericardial heterografts, preserved dura and the polytetrafluoroethylene patch (PTFE),have been advocated. The most commonly used prosthetic material presently is Surgisis soft tissue graft, which is incorporated into adjacent tissue, and this tends to lessen the risk, extension or displacement, with a decreased risk of infection. Abdomen is closed in layers. If the abdominal cavity is small, gentle stretching of the abdominal wall will enable safe closure in most of the patients. Chest drain should be avoided. The argument against the use of a chest drain is in avoidance of barotraumas as it increases the transpulmonary pressure gradient.

Plication of the diaphragm has been used for many years to treat eventration. Plication increases both tidal volume and maximal breathing capacity and has been successful in many clinical series. An abdominal approach through a subcostal incision is preferred for left-sided eventration but a thoracic approach through a posterolateral incision via the sixth space may be used for right-sided lesions. The transabdominal approach allows good visualization of the entire diaphragm from front to back and easier mobilization of abdominal contents.

Postoperative care.

Although the diaphragmatic hernia can usually be repaired, if both lungs are markedly hypoplastic, adequate oxygenation will never be achieved. Survival may be compromised in babies with relatively good lungs if pulmonary artery hypertension occurs. Extracorporeal

membrane oxygenation can decrease the degree of pulmonary artery hypertension by producing adequate oxygenation, alkalosis, and hypocarbia. All three factors will lower pulmonary artery pressures, and this technique may save some infants who previously have succumbed to persistent fetal circulation.

Ventilatory support is nearly always needed following repair. It must be regulated to limit inspiratory pressures and minimize barotrauma to the lungs. The hypoplastic lung bud should not be distended artificially. Close monitoring of arterial blood gases is essential: deterioration often indicates rising pulmonary artery pressures and additional sedation with fentanyl should be instituted. Deliberate hyperventilation to produce alkalosis and hypocarbia will decrease pulmonary artery resistance and hence pressures. Comparisons of preductal (right radial artery) and postductal (umbilical artery or posterior tibial artery) gases will define the extent of shunting at the ductal level.

Complication. Acute deterioration may be due to a pneumothorax on the contralateral side: this should be confirmed radiographically and treated with intercostal tube drainage.

Further inpatient care. Babies may require several weeks of hospitalization after surgery depending on how long breathing needs to be supported with a machine. Feeding is begun after the first bowel movement is passed. Feeding is usually done through a tube into the stomach or small intestines until the breathing tube is removed.

Prognosis. Isolated diaphragmatic hernia is an anatomically simple defect, which is easily correctable, the mortality rate is about 50%. The main cause of death is hypoxemia due to pulmonary hypertension, resulting from the abnormal development of the pulmonary vascular bed.

5. Additional materials for the self-control

A. Clinical cases

Case 1. In a 12 hour new-born 3-4 hours after birth there appeared foamy saliva, the attacks of shortness of breath and vomiting with milk appear at feeding. Meconium stool is in small amounts.

What diagnosis have you made?

What auxiliary methods of examination are to be used?

Case 2. Since birth a 5 day-old boy has shortness of breath, dry cough, which intensifies at feeding, and the child turns blue after that. During percussion in the left part of thorax a tympanic sound is heard, the heart is displaced to the right. During auscultation on the left weak breathing, and intestinal murmurs are heard. The abdomen is hollow.

What diagnosis have you made?

What auxiliary methods of examination are to be used?

What tactics have you developed?

Case 3. In a new-born child in 6 hours after birth there appeared gradually incresing the respiratory impairment - shortness of breath, cyanosys. At the repeated examination gradual displacement of heart to the right is detected .The left half of thorax somehow protrudes, falls behind in the act of breathing. During percussion on the right there is an ordinary pulmonary sound, on the left there is shortening of the pulmonary sound, tympanitis appears periodically. During auscultation on the left to the "gurgling" murmurs are hearkened. On the survey sciagram of the thorax organs mediastinum is displaced to the right, on the left to the second rib air cavities of different size are identified.

What diagnosis have you made?

What auxiliary methods of examination are to be used?

What tactics have you developed?

B. Tests

6.LITERATURE FOR STUDENTS

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