“ ЗАТВЕРДЖЕНО”

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професор\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_А.Ф. Левицький

**Theme № 1. Congenital Malformations of Respiratory System in Children**

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**Overview:**

Congenital anomalies account for one third of infant deaths and are one of the leading causes of death in this age group in most developed countries. Congenital malformations of the respiratory system now rank second, behind those of the cardiovascular system, as a cause of infant mortality. With a rate of 0.25 death per 1000 live births, they have surpassed those of the nervous system (0.23 per 1000) in the past decade in the United States. However, congenital malformations of the lungs and airways include a wide spectrum of developmental anomalies, some of which remain asymptomatic and are discovered incidentally on imaging studies. Their frequency has been reported to range from 7.5% to 18.7% of all congenital malformations, 4,5 but their exact overall incidence is difficult to ascertain. They may be part of more complex syndromes and are often associated with other congenital anomalies, particularly those involving the heart and great vessels.

In the structure of morbidity of the population of Ukraine respiratory diseases traditionally occupy 1st place in children, 3rd place in infant mortality that comprise about 12%, and in the structure of disability they take 4th place. Anomalies of the tracheobronchial tree and lungs found in about 10% of children with the syndrome of airway obstruction.

**Educational aims:**

The aim of this part of module is to provide help in identifying those children with congenital malformations of airway and lungs and to provide guidance on the diagnosis, differential diagnosis, defining indications for surgery and choice of optimal surgical treatment.

**A student must know:**

1. Relevant anatomy of the airway, lungs and mediastinum in children.

2. Basic embryology of respiratory system

3. Nomenclature of malformations presenting with respiratory insufficiency

4. Main clinical signs characterizing the malformations of airway, lungs, esophagus, diaphragm that present with respiratory distress.

5. Diagnostic modalities used in infants and children with malformations of respiratory system

6. Main radiological signs of individual congenital malformations of airway, lungs, esophagus, diaphragm.

7. Main endoscopic signs of individual congenital malformations of airway.

8. Differential diagnosis of individual congenital lesions of airway, lungs, esophageal atresia and congenital diaphragmatic hernia.

9. Types of congenital airway stenoses, their clinical presentation, diagnosis and treatment.

10. Variants of vascular rings causing compression of the airway, their clinical presentation, diagnosis and treatment.

11. Clinical presentation, diagnosis and management of bronchogenic cysts.

12. Clinical presentation, diagnosis and management of pulmonary aplasia, agenesis and hypoplasia.

13. Clinical presentation, diagnosis and management of congenital emphysema.

14. Clinical presentation, diagnosis and management of cystic lung lesions.

15. Clinical presentation, diagnosis and management of pulmonary sequestrations.

16. Clinical presentation, diagnosis, prenatal diagnosis and management of congenital diaphragmatic hernia.

17. Clinical presentation, diagnosis and management of esophageal atresia.

**A student must be able to**

1. To collect complaints and history in patients suspicious for malformations respiratory system and their parents.

2. To determine the clinical signs of respiratory failure

3. To prescribe laboratory and instrumental examination of patients with symptoms of respiratory disorders.

4. To evaluate the results of X-ray observation in patients with malformations of respiratory system.

5. Describe the technique esophagogram, determine the indications for its use and evaluate the results.

6. Evaluate the results of computed tomography in patients with congenital malformations of respiratory system, determine the indications for its use.

7. Describe the technique of bronchoscopy and its features and value in patients with malformations of respiratory system.

8. Evaluate the results of prenatal ultrasound diagnosis of malformations of the respiratory system of the fetus.

**Terminology**

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| **Term**  | **Definition** |
| Norma  | complete absence of anatomic divergence from the classic description of organ’s structure with full function of all of the structural units |
| Anomaly  | anatomic changes of an organ, that under ordinary conditions do not cause violation of its function, but with the development of pathological process, it may cause influence on its function |
| Malformation  | rough morphological changes in an organ or system with violation of their function |
| Respiratory insufficiency  | The pathological condition in which normal level of blood gases (Pa O2 - 92 - 100 mm Hg .; Pa CO2 - 35 - 45 mmHg) is provided (or not provided) due to exertion of compensatory mechanisms that significantly affects the patient's condition |
| Stridor  | noisy breathing, which occurs when the airway lumen is narrowed by 2/3 or more at the level of the larynx, trachea or main bronchi |
| Respiratory distress-syndrome | In a narrow sense - respiratory distress syndrome in newborns is associated with surfactant deficiency;In a wider sense it means respiratory disorders and respiratory failure associated with pulmonary injury or violation of airway patency |

**Content**

**CONGENITAL AIRWAY MALFORMATIONS**

The group of congenital malformations of the airway, that presents with respiratory distress include large amount of following entities: choanal atresia, laryngomalacia, laryngeal web, atresia and stenosis, Subglottic hemangioma, tracheal stenosis, bronchogenic an enterogenic cysts, etc. Most of them cause airway narrowing and obstruction followed by respiratory failure even since the first days of life. Traditionally, defects of the upper respiratory tract (the level of the beginning of trachea) are within the competence of ENT doctors, but in terms of diagnosis, differential diagnosis and timely medical care they certainly are interesting for the doctor of any specialty.

**Choanal atresia** is a congenital obstruction of posterior nasal passages, may be membranous or osteal, unilateral or bilateral. Unilateral choanal atresia. In the case of bidirectional choanal atresia nasal breathing becomes impossible, the child breathes through the mouth. Apnoic attacs occur during sleep or feeding. Diagnosis is made by the attempts of passing the catheter through the nose into the nasopharynx. Contrast study and nasopharyngoscopy may be useful. The baby is fed through the nasogastric tube. The treatment is surgical. The early correction is recommended. In the cases of impossibility of adequate ventilation the surgery is indicated in the first days of life. The cutoff of septum is made through the palatal or nasal approach depending on age of the child.

**Laryngomalacia** is the most common cause of congenital stridor in infants, accounting for approximately 60% (range 50–75%) of all congenital laryngeal anomalies. Boys are affected twice as often as girls. Laryngomalacia is an enigmatic disease of unknown etiology. It is believed to be due to a delay in maturation of the supporting laryngeal cartilages, causing an inward collapse of the supraglottic structures on inspiration. A high-pitched fluttering inspiratory stridor is the hallmark of laryngomalacia. Typically, the stridor worsens during increased airway demands, such as crying, feeding, or the child’s being in the supine position. Usually, the course of the disease is self-limiting, with onset around the age of 2–4 weeks, progression to a culminating point at around 6–8 months, and resolution occurring by 2 years of age. Feeding difficulties are related to gastro-esophageal reflux in up to 80% of cases. Regurgitation, recurrent vomiting, occasional coughing, or choking are seen in moderate to severe cases. Aspiration is often due to uncoordinated breathing and swallowing during deglutition. In 5 - 10% of cases laryngomalacia can cause apnea, feeding difficulties and cor pulmonale. These patients require surgical correction. Mild cases of laryngomalacia, seen in 80–90% of infants, require only diagnostic confirmation conducted in the outpatient clinic by awake transnasal fibreoptic laryngoscopy. In most cases such patients do not require special treatment. A symptomatic therapy is administrated. Prednisone (2 mg / kg) is assigned for 10 - 14 days. Patients with severe and progressive respiratory insufficiency may require endotracheal intubation or tracheostomy. Over the past 20 years endoscopic intervention, namely CO2 laser epiglottoplasty or supraglottoplasty have become spread widely. The goal of these procedures is to widen the supraglottic lumen and thus to overcome the inspiratory obstruction. The operation includes the vaporization of excessive mucosa of arytenoid folds and trimming the epiglottis. Efficiency of supraglottoplasty varies between 80 - 100% according to various studies.

**Congenital laryngeal atresia, webs, and stenosis.** *Laryngeal atresia* is a life-threatening malformation. The lesion results from failure of recanalization of the epithelial septum, which forms around 6 weeks of gestation and separates the developing esophagus from the tracheal bud. In the past, affected babies would often die at birth or have severe neurologic sequelae because of the failure to establish an airway. In recent years, routine prenatal ultrasound examinations have allowed the diagnosis to be made before birth, resulting in a higher survival. On ultrasound, the lungs appear more echogenic and enlarged, causing eversion of the diaphragms; a distended trachea can also be observed. Magnetic resonance imaging confirms the increased fluid content of the lung, with a hyperintense signal on T2-weighed images. Near term, the fetus is delivered using the EXIT procedure (EXutero Intrapartum Treatment), which is a cesarean section done under deep maternal general anesthesia to provide uterine relaxation. The fetal head and neck are exposed and the airway is secured, usually by tracheostomy, while oxygenation is maintained through the placenta.

The prognosis is better with *laryngeal webs*, which can present with openings of varying sizes. Of these, 75% are glottic, with the rest supraglottic or subglottic in location. Complete webs present like atresias, whereas partial webs present with stridor and a hoarse or weak cry and may cause varying degrees of respiratory difficulty depending on the degree of obstruction. Partial webs may also present as a difficult to intubate neonate who is placed under general anesthesia for repair of another malformation such as esophageal atresia. The diagnosis of laryngeal webs is confi rmed at endoscopy and treatment is excision or laser ablation, although some smaller subglottic webs may respond to dilation.

The **congenital subglottic stenosis**, similar to laryngeal web and atresia, results from defective recanalization of the larynx, although in subglottic stenosis the defect occurs at the level of, and usually involves, the cricoid cartilage, approximately 2 to 3 mm below the glottis. The most common presenting symptom is stridor, which is worsened by increased respiratory effort and upper respiratory tract infection. In fact, in milder forms the stridor may only be noticed during intercurrent upper respiratory tract infections and be mislabeled as croup. Therefore, recurrent “croup” in infancy should always raise the possibility of a fixed upper airway narrowing such as subglottic stenosis, particularly when the course of the illness and response to treatment are atypical. The diagnosis is made by using high-voltage radiographs of the upper airway in anteroposterior and lateral views and flexible endoscopy. Bronchoscopy is the definitive diagnostic tool and is usually necessary to exclude other causes of narrowing in this region, particularly subglottic hemangioma. Acquired subglottic stenosis is much more common than the congenital form. The diagnosis of the former is supported by a history of laryngeal trauma, the most common of which would be endotracheal intubation. This is particularly true when intubation is prolonged, as in the premature neonate. Because the subglottic narrowing generally improves with laryngeal growth, a conservative approach using supportive care, particularly during intermittent episodes of “croup,” should be the goal in all patients. Surgery should be reserved for patients who fail to cope with this conservative management, and a cricoid split is often the first line surgical treatment. cricoid split procedure, laryngotracheoplasty, tracheal resection and anastomosis, and tracheostomy for patients who have lesions that are not amenable to resection or reconstructive procedures. Most tracheostomized patients can be successfully decannulated within 2 to 3 years, although stridor and varying degrees of breathing difficulties may persist for many years.

**Subglottic hemangioma** is a rare benign tumor of the airway, accounting for only 1.5% of all congenital laryngeal anomalies. Subglottic haemangioma is more common in female than in male patients with a ratio of 2–3:1 and are potentially lifethreatening in the absence of treatment. They are benign tumors associated with hyperplasia of the endothelial cells, mast cells, pericytes, fibroblasts and macrophages. In contrast, vascular malformations display a normal cell turnover rate. The severity of the disease depends on the size of the tumor. Respiratory viral infections and restlessness enhance respiratory disorders. Understanding of evolution of subglottic hemangioma is essential in making a decision about the best treatment option. They undergo a rapid proliferative phase lasting a few months, followed by a period of stabilisation, and finally, a slow involution phase of several years. According to A.L. Bruckner (2006), complete resolution occurs in 50% of the cases by the age of 5 years, 70% by the age of 7 years and 100% by the age of 10–12 years. Due to this typical evolution pattern, the clinical course is often stereotyped even if there are wide variations in rate, degree and duration of the disease. During the first weeks of life, the infant is asymptomatic. Usually, symptoms of inspiratory stridor followed by biphasic stridor with barking cough and slight hoarseness start at around 2–4 months, becoming manifest in all infants by the age of 6 months. Symptoms of respiratory distress with suprasternal and chest retractions, feeding difficulties and failure to thrive depend on the severity of the airway obstruction. The progression of symptoms reaches a plateau between the ages of 10 and 12 months, and the symptoms then decrease slowly and finally disappear around the age of 2 years, although complete resolution of the tumor may take as long as 5–10 years. Based on the clinical history and examination, this presentation must be differentiated from a subglottic cyst, stenosis or papilloma. The mainstay of the diagnosis is rigid endoscopy under general anesthesia after laryngomalacia and vocal cord paralysis have been ruled out by fiber optic laryngoscopy. An extensive or therapy-resistant large hemangioma warrants a contrast-enhanced MRI or CT scan to identify potential extension into the upper mediastinum. The subglottic hemangioma appears as a reddish smooth mass, mostly located in the subglottis and extending cranially to the under surface of the vocal cord. The tumor mass is spongy and compressible, allowing for easy intubation with an ET tube with no risk of major hemorrhage. Knowing that subglottic hemangioma is a self-limiting disease with spontaneous resolution, treatment must be aimed at maintaining the airway without tracheotomy, while avoiding any long-term sequelae. Treatment modalities include observation, medical treatment (systemic steroids, propranolol), endoscopic treatment (intralesional steroid injections, laser resection) and open surgery (tracheostomy, excision through a laryngofissure or tracheotomy, tracheal resection with anastomosis). Observation is appropriate for children with mild symptoms as well as children older than 1 year having reached the phase of spontaneous tumor regression. Systemic steroids (prednisolone 4-5 mg/kg per day) should be used as adjuvant therapy or as curative treatment only for a short period. Clinical efficiency of this treatment if administered alone is approximately only 25%. Boston protocols do not recommend systemic steroid treatment for longer than 3 weeks if the symptoms do not markedly improve. Potential long-term side effects such as failure to thrive, osteoporosis, adrenal suppression and Cushing syndrome should not be underestimated. Administration of propranolol is rather new and efficient method of treatment of hemangiomas, and subglottic or tracheal hemangioma in particular. Possible explanations for the therapeutic effects of propranolol include vasoconstriction, decreased expression of VEGF and βFGF genes via the downregulation of the RAF-mitogen-activated protein kinase pathways, as well as the triggering of apoptosis of capillary endothelial cells. Propranolol is used at 2 mg/kg of bodyweight per day during at least 6 months. Endoscopic intralesional steroid injections is reported to have success rates exceeding 75%, this treatment modality is much more efficacious than systemic steroids. Laser resection is appropriate for slow-growing tumors that become symptomatic at the age of 4–6 months. Open surgery was found to be the most efficient treatment, albeit with potential complications. It is mostly suitable for fast-growing tumors or bilateral tumors. In the future, propranolol might well supersede all other treatment modalities for children who do not have a contra-indication to the prescription of b-blocking agents.

**Laryngeal cysts.** These are usually supraglottic and generally present in the neonatal period (although possibly much later) with hoarse or muffled voice or even aphonia, stridor, and respiratory difficulty. A lateral neck radiograph may show a rounded supraglottic swelling, and at laryngoscopy, a bluish fluid-filled cyst is found, usually in the epiglottic folds. Aspiration can relieve acute symptoms, but resection is ultimately necessary to prevent recurrence.

**Tracheal stenoses**

The narrowing of the trachea are divided into congenital and acquired. Congenital tracheal stenosis have different morphological reasons and distributed to intrinsic tracheal stenosis (complete cartilaginous rings), compression stenosis (due to the anomalies of great vessels - congenital vascular ring or mediastinal tumors and cysts) and functional stenosis (tracheomalacia). Acquired stenosis may be of compression (mediastinal tumor) or cicatricial nature. The former result from prolonged intubation, tracheostomy, foreign bodies, burns and traumatic ruptures of the respiratory tract. Congenital intrinsic tracheal stenosis is usually associated with complete cartilaginous tracheal rings and absence of tracheal membrane causing narrowing of tracheal lumen varying in lengths. A number of forms have been recognized. Cantrell J.R. and Guild H.G. (1964) based on 24 observations of morphological variants identified three types of congenital tracheal stenosis: generalized hypoplasia (30%), funnel-shape stenosis (20%), segmental stenosis (50%). This classification was later modified by H.C. Grillo fnd the 4th type was described.

Type I: generalised tracheal hypoplasia. Almost the entire trachea is stenotic while only the first to third cranial rings are normal.

Type II: funnel-shaped tracheal narrowing. The abnormal tracheal segment varies by location and length, but always has a funnel configuration that is shaped from the cranial to caudal direction.

Type III: segmental tracheal stenosis. This type is characterised by a short-segment stenosis located at different levels of the trachea, at times below an anomalous right upper lobe bronchus.

Type IV: bridge bronchus stenosis. In this variant of Type III, the anomalous right upper lobe bronchus is in the proximity of the carina, and via horizontally branching bronchi, the stenotic bridge bronchus connects the proximal trachea to the rest of the lungs.

In general, the onset of symptoms starts a few months after birth when the baby’s activities lead to an increase in respiratory demands. The length is less critical than the degree of the stenosis in producing symptoms. Biphasic stridor with predominant wheezing, chest retractions, cyanotic attacks and recurrent pneumonia are likely to occur during the first respiratory illness. When the narrowing is severe, breath sounds may be accompanied by significant respiratory distress, whereas in mild cases they may be noticeable only when respiratory load is increased, as with exercise or infection. Depending on the degree of stenosis a defect may be compatible with long life. However, in such patients inflammatory complications in the lung, bronchiectasis, pulmonary hypertension and pneumofibrosis join over time accompanied with progressing of respiratory and cardiovascular failure. Penetrated radiographs of the airway may identify a narrowed segment, although CT or MRI is necessary to define the extent of the lesion and to search for an extrinsic cause. CT scan with 3D reconstructions and an MRI with digital subtraction are the examinations of first choice for achieving an accurate diagnosis. These tools permit a clear assessment of the relationship between the airway and mediastinal cardiovascular anomalies if present. Bronchoscopy may also be helpful to plan treatment. The endoscopist must be careful to prevent trauma during the examination. In fact, the slightest mucosal injury is likely to decompensate the compromised airway, provoking an acute airway crisis. In the absence of infection, tracheal cartilages are readily visible during endoscopy and must be precisely counted. Normal rings are easily distinguished from circular rings. Respiratory function tests show evidence of fixed obstruction with characteristic flattening of the inspiratory and expiratory portions of the flow-volume loop. Management is difficult. In some patients the stenosis improves with tracheal growth, and conservative symptomatic treatment and support should be the recommended approach when possible. Results of dilation techniques and laser resection of the intraluminal narrowing have been disappointing, with subsequent recurrence of the stenosis the rule. Endoscopic stenting has been used in some centers. This should be avoided when there is an associated vascular ring or sling because of the risk of erosion with arterial fistulization. Five main open surgical approaches have been used: resection with end-to-end anastomosis, enlargement patch tracheoplasties (pericardial, cartilage), tracheal autograft technique, slide tracheoplasty, tracheoplasty with cadaveric tracheal homografts. Primary resection with end-to-end anastomosis is suitable for short segment stenoses not exceeding one-third of the tracheal length. Slide tracheoplasty is now the technique of choice for long-segment tracheal stenosis as it fulfils the basic requirements for adequate airway reconstruction, that is steady cartilaginous support and a fully mucosalised inner lumen. The basic principle consists of doubling the tracheal circumference at the level of the stenotic segment. This is achieved by transecting the stenosis horizontally at its midpoint, slitting the upper and lower stenotic segments anteriorly and posteriorly, respectively, and then sliding the two segments over one another. Use of cardiopulmonary bypass provides better conditions for the operation as it secure stable gas exchange at the period of opened airway. Tissue engineering will undoubtedly provide a useful alternative for tracheal reconstruction in the future.

**Compression tracheal stenoses** may be caused by malformations of the aortic arch and its vessels, pulmonary artery (vascular rings and loops), and congenital cysts and tumors of the mediastinum (teratoma, hemangioma, bronchogenic cysts).

**Vascular rings and slings**

Abnormally developed aortic arch and its branches and pulmonary artery form complete or incomplete vascular ring around the trachea and esophagus and cause their compression and severe airway obstruction resulting in respiratory disorders that may be life-threatening. Vascular rings do not cause cardiovascular symptoms, so usually drop out of sight of cardiac surgeons. Their clinical significance relates entirely to the effects of compression of the trachea and esophagus. Prolonged inadequate treatment of children in somatic hospitals without timely surgical care threatens death from asphyxia or chronic inflammation in the airways and lungs of patients with disability.

Vascular rings are uncommon malformations. Anomalies of the aortic arch and its branches are 0.5 - 3.5% of all congenital cardiovascular system. Most vascular ring is found in isolated form. There is an association with patent ductus arteriosus or septal defects of the heart, Fallot’s tetralogy, pulmonary atresia, and transposition of the great arteries.

Most vascular rings are associated with varying degrees of persistence of bilateral fourth aortic arches during early development. The model of hypothetic double aortic arch by Edwards (1948) consists of right and left aortic arches, two ductus arteriosus and two upper descending aortas. The normal aortic arch is the result of regression of the right upper descending aorta and the right ductus arteriosus.

Anatomical variants of vascular rings and slings are following

1 - double aortic arch;

2 – right aortic arch with left ligamentum arteriosum

3 - aberrant right subclavian artery;

4 - pulmonary artery sling;

5 – innominate artery compression

Various malformations of this group are united by common embryogenetic basis and tracheoesophageal compression syndrome.

**Double aortic arch** (DAA) is the most common variant of the vascular ring and occurs in about 50% of cases. It is caused by the persistence of the embryological fourth aortic arch creating a complete ring around the trachea and esophagus. Each arch gives rise to a carotid and subclavian artery and the descending thoracic aorta may be located leftward or rightward side of the vertebral column or take midline position. In 45–77% of cases, the aorta descends on the left side. Based on the patency of aortic arches the balanced arches, right arch dominant and left arch dominant forms of anomaly are distinguished. In the majority of cases, the right arch is the larger. Rarely, there is an atretic arch (left arch in most cases). The separate branching of brachiocephalic vessels is common without brachiocephalic trunk formation.

The first report on DAA was made by Hommel in 1737, in 1932 Maude Abbott described 5 postmortem cases of DAA considering the possibilities of surgical treatment. In 1939, Wolman reported the syndrome of esophageal and tracheal compression caused by a double aortic arch and a few years later, in 1945, Gross had successfully repaired this type of lesion, followed by the correction of many other forms of vascular rings. In Ukraine for the first time the malformation was diagnosed and successfully surgically corrected in 1 year-old girl by prof. D.U. Krivchenya in 1982.

**Right aortic arch with left-sided ductus**. This is the second commonest vascular ring (25–30%). The characteristic features of anomaly are as follows: ascending aorta takes its normal position, aortic arch is located to the right side of trachea and esophagus and passes over the right main bronchus; descending aorta may take either right-sided or left sided position; ductal ligament which connects left pulmonary artery and aortic isthmus closes complete vascular ring that compresses the trachea and esophagus. The aortal diverticulum (Kommerell’s) may be present at the site of aortal end of ligamentum arteriosum which more often seen in the cases of right-sided descending aorta and enhances the posterior compression of esophagus and trachea. The aberrant left subclavian artery often arise from this diverticulum.

There are two main variants of the malformation: right aortic arch with aberrant left subclavian artery and right aortic arch with mirror image branching of brachiocephalic vessels. In the former type left common carotid and left subclavian arteries originate from the aortic arch together forming left brachiocephalic trunk.

Thus, the complete vascular ring in this anomaly is formed by the aortic arch on the right side and on back, including diverticulum of Kommerell, ligamentum arteriosum on the left side and pulmonary artery on front.

Double aortic arch and right aortic arch are most commonly found isolately, but association with patent ductus arteriosus is possible. The rare cases of combination with coarctation of aorta and pulmonary artery sling are reported. According to A. Hastreiter et al (1966), right aortic arch with mirror image branching of brachiocephalic vessels is always combined with congenital heart defects.

**Aberrant right subclavian artery** is the most common anomaly of the aortic arch, appearing in 0.5–0.7% of autopsies. The right subclavian artery arises from the descending aorta and crosses to the right side, mostly behind the esophagus. This anomaly results in the regression of a segment of the ‘hypothetic double aortic arch’ between the right common carotid artery and the right subclavian artery. In this anomaly vascular ring is incomplete (semi-ring or sling). There are 4 vessels arising from the aortic arch separately: right and left common carotid arteries, left subclavian artery and, at last, right subclavian artery which arises from the distal part of aortic arch. The incomplete ring around the trachea and esophagus is formed by aortic arch on front and aberrant right subclavian artery on back which on its way to right arm causes posterior compression of esophagus and trachea.

**Pulmonary artery sling** is characterized by abnormal development of bifurcation of the pulmonary artery trunk. The sling is created by anomalous origin of the left pulmonary artery, which arises as the first branch of the right pulmonary artery. The vessel runs between the trachea and esophagus toward the left hilum. The trachea is trapped in the sling created by the left pulmonary artery. A ductal ligament running from the main pulmonary artery completes the ring. This looping of the aberrant vessel around the carina usually has a strangling effect, resulting in variable degrees of compression and localized narrowing. Thus, the compressive stenosis of the trachea at suprabifurcation area is formed.

There are two forms of anomaly: with normal anatomy of tracheobronchial anatomy and with airway malformation (abnormal bronchial branching at bifurcation in the area, tracheomalacia and complete cartilaginous rings). Combination of pulmonary artery sling with tracheal stenosis due to complete cartilaginous rings was named as "ring-sling complex". Its frequency is up to 50 – 60%. The complex morphological features of the malformation are complemented by pathology of lung vessels. The disorders of lung ventilation is enhanced by compromised blood circulation in the lungs.

Description for the first case of pulmonary artery sling was made by Glaevecke and Doehle in 1897, as a autopsy finding in 7-month baby suffering from severe respiratory distress. The malformation is rare and highly lethal. If untreated 90% of children die within first months of life. Postoperative mortality varies between 50% in early reports and 5% nowadays. Since the first report in the literature by 1995, about 150 cases of anomaly have been reported in the literature and about half of them were clinical observation, the other half autopsies. Associated severe cardiovascular defects occur in 30-50% of cases.

**Abnormal brachiocephalic trunk** is the most common cause of vascular compression of the trachea. It is also known as innominate artery compression syndrome. Tracheal compression occurs due to a relatively distal branching of the innominate artery which then crosses the trachea causing its anterior compression in association with a narrow thoracic inlet. First description was given by R.E. Gross and E.B.D. Neuhauser in 1948. Another anatomic variant of the anomaly is trifurcation of brachiocephalic trunk when it divides into three vessels: the right subclavian, right and left common carotid arteries. In this case brachiocephalic trunk has a larger diameter and also originates more distally from the aortic arch, causing anterior tracheal compression. Prolonged compression of tracheal wall results in the disruption of the cartilage structure of airway - secondary tracheomalacia.

The **clinical picture** of compression tracheal stenoses depends on the degree of narrowing of the trachea and esophagus and presents with respiratory and esophageal groups of symptoms. The existence and severity of symptoms are related to the tightness and completeness of the vascular ring. Respiratory signs include inspiratory stridor with or without an association with an expiratory wheeze and tachypnea. The commonest presenting symptom of a vascular ring is stridor. Around 50–70% of symptomatic cases present during the first year of life. Various positions, such as lying on the back, may aggravate the stridor, while it may be relieved by extension of the neck. Other signs are noisy breathing, a hoarse cry and persistence of a barking cough. In severe cases there are episodes of apnea with cyanosis and unconsciousness. Recurrent respiratory infections aggravate the condition and are more frequent in vascular rings. In mild tracheal compression, symptoms may be present only at the time of respiratory infections. As a result of esophageal compression, the baby feeds poorly and its growth may be retarded. There is difficulty in swallowing liquids and solids. Regurgitation and choking, with aggravation of respiratory symptoms, are common in severe cases, whereas in mild cases of obstruction, dysphagia may be the only symptom.

**The natural course vascular rings.** Symptoms can be severe from the first days of life and may become critical in the first few months. Following the violation of ventilation due to tracheal compression airway infection, delay sputum evacuation, pneumonia and lung hyperinflation occur. With the increase in the volume and density of food the aspiration syndrome develops due to compression of the esophagus. In this vicious circle there is no hope of survival of patient without surgery. Medical therapy is usually ineffective, provides temporary relief of symptoms. In most cases lethal outcome occurs at age 1 - 2 years from pulmonary complications.

**Diagnosis.** The examination begins with plain chest X-ray, although the method does not show direct signs of compression. A chest radiograph is mandatory in any symptomatic child. The plain X-ray shows prominence of the mediastinum to the right side in double aortic arch, right aortic arch with aberrant left subclavian artery, right aortic arch with mirror-image branching, and left ductus arteriosus. The aortic knob is on the left side in cases of left aortic arch with aberrant innominate artery, left aortic arch with aberrant right subclavian artery (in this case the aberrant vessel may be seen above the aortic knob). Indirect signs of obstruction is emphysema (single or bilateral), atelectasis and secondary inflammatory changes in the lungs.

The most useful first-line investigation is esophagography , i.e. contrast radiography of the esophagus in the anteroposterior and lateral projections. This is highly informative, easy and safe carrying study, which does not injure the respiratory tract exhausted in terms of compensation. The presence of characteristic filling defects of esophagus points to a compression of the esophagus and the trachea. Frontal esophagogram shows impression both on the right and left sides in cases of double aortic arch, right aortic arch with mirror-image branching and left ductus arteriosus. Oblique filling defect from the right, going upwards to the left, is seen in aberrant left subclavian artery with a right aortic arch. In pulmonary artery sling, the esophagus may be deviated to the left. Lateral esophagogram shows posterior compression in cases of double aortic arch, aberrant right subclavian artery, right aortic arch with mirror-image branching and left ductus arteriosus. In pulmonary aortic sling, the esophagus is compressed from the front. In anomalous origin of innominate artery there is no compression on the esophagus but the study is advisable to exclude other variants of vascular ring.

Aortography in axial projection is of optimal informativity in the cases of double aortic arch, right aortic arch with left ligamentum, aberrant right subclavian artery and abnormalities of brachiocephalic trunk, and selective pulmonary arteriograghy is optimal for diagnosis of pulmonary artery sling

However, angiography has been largely superseded by CT/MRI although occasionally this may be necessary if there is doubt about which side of the arch is dominant. Ligamentous structures (i.e., a ductus ligament or the ligamentous remnant of a double arch) cannot be seen on either echocardiography or CT scan but their position can be inferred from the nature of the ring. Computerized tomography, with intravenous contrast, or MR scanning are required to define the vascular anatomy in detail. Increasingly MR with 3-D reconstruction of the vascular structures and trachea is becoming the preferred method of imaging vascular rings.

Echocardiography should also be performed in order to exclude associated cardiac anomalies and to complete the diagnosis.

Bronchoscopy is not essential as a part of routine assessment of a vascular ring unless a pulmonary artery sling is suspected. However, some infants and children with vascular rings will have had airway endoscopy because they present with stridor. The endoscopic appearances are characteristic, with pulsatile extrinsic compression of the airway. In the area of compression tracheal cartilage are often not visualized that is the sign of secondary tracheomalacia. Bronchoscopy could cause injury to mucosa and to increased obstruction due to swelling of the mucous membranes, so it should be done immediately before surgery. Airway examination by bronchoscopy does form an important part of the assessment of a pulmonary artery sling because of the common association with tracheal stenosis.

**Differential diagnosis.** Because most children with vascular rings present with stridor, the differential diagnosis includes other causes of stridor. The history can provide important clues – for example, the acute presentation of laryngotracheobronchitis or epiglottitis is totally different from the chronic history of a vascular ring. Laryngomalacia is a common cause of stridor in infancy. The stridor is high-pitched, musical, and exclusively inspiratory. If there is doubt about the diagnosis airway endoscopy and/or a barium swallow will clarify matters. Subglottic stenosis is usually acquired as a consequence of prolonged endotracheal intubation and, therefore, more common in ex-premature infants. The diagnosis is made by airway endoscopy.

It is necessary to conduct differential diagnostics of compression syndrome caused by tumors and cysts of the mediastinum, including bronchogenic and enterogenic cysts, esophageal diverticulum.

Differential diagnosis between vascular ring variants is important to select the optimal method of surgery.

**Treatment** of vascular compression stenosis of the trachea is only surgical. The essence of the operation consists of division of vascular ring and depends on anatomic variant of the anomaly. In the case of double aortic arch functionally less significant or atretic aortic arch is divided as well as ductal ligament (or patent ductus arteriosus). In the case of secondary tracheomalacia the operation is complemented with anterior or posterior aortopexy to stabilize softened trachea. Correction of right aortic arch with left ligament include division of the ductal ligament and, if necessary, oposterior aortopexy or resection of Kommerell’s diverticulum. Compression caused by aberrant right subclavian artery is corrected by means of division of this vessel at the site of its branching from the aortic arch. Circulation of the upper limb is kept by the presence of natural collaterals. However, some surgeons recommend reimplantation of the right subclavian artery into the right common carotid artery, thus forming the normal brachiocephalic trunk.

Correction of pulmonary artery sling is a complex reconstructive surgery that may require cardiopulmonary bypass. The left pulmonary artery is dissected as far as possible and freed from the adjacent structures while the ligamentum arteriosum is divided. The pulmonary artery is divided between clamps and the proximal stump closed. The left pulmonary artery is then retrieved from behind the trachea and brought forward to assume the anatomical position next to the main pulmonary artery. The left pulmonary artery is anastomosed to the main pulmonary artery and to the side by using a side-biting clamp. If the pulmonary artery sling is associated with tracheal stenosis this should be repaired at the same time. Localized stenoses can be treated with resection and reanastomosis. A sliding tracheoplasty may be required for longer stenoses. More extensive stenoses with complete tracheal rings can require complex reconstruction on cardiopulmonary bypass and should only be undertaken by specialist teams.

Anomalous origin of the innominate artery is usually treated by suspension of the proximal part of the artery and the aorta to the sternum. The innominate artery can be approached via mid-sternotomy, right anterior thoracotomy or left anterior thoracotomy. The anterior ligament aortopexy first proposed by prof. D.U. Krivchenya allows avoiding such complications as suture tearing, bleeding and recurrence of airway compression.

Surgical correction of all variants of vascular rings for the first time in Ukraine developed and implemented by prof. D.U. Krivchenya during 1982 – 1989.

**Bronchogenic and enterogenic cysts**

Benign congenital cystic lesions have been referred to by various names, including bronchogenic cyst, esophageal duplication cyst, enteric cyst, and neuroenteric cyst. They are all believed to constitute congenital abnormalities of division of the embryonic primitive foregut.

Bronchogenic cysts are the most common of the bronchopulmonary malformations encountered in children. These lesions are usually unilocular, are lined by ciliated columnar epithelium and contain fragments of cartilage and smooth muscle within the wall. Bronchogenic cysts usually demonstrate no patent communication to the airway but are almost always closely attached to the major airways or to the esophagus by dense fibrous tissue.

Enteric cysts of the mediastinum contain gastric mucosa and may or may not communicate with the gastrointestinal tract below the diaphragm. These cysts may be associated with vertebral anomalies, but there exist no intervertebral communication.

Neuroenteric cysts contain both endodermal and ectodermal, or neurogenic, elements. Characteristically, a neuroenteric cyst is connected by a stalk to the meninges and spinal cord. They are associated with spinal deformities, including congenital scoliosis, hemivertebrae, and spina bifida.

In contrast to the situation in adults where these lesions are incidental findings, in children these cysts are usually symptomatic and sometimes life-threatening. Symptoms include persistent cough, progressive dyspnea, wheeze, stridor, and cyanosis. Additionally, gastroenteric cysts might be associated with pain or melena from peptic complications secondary to the activity of the gastric tissue.

Plain chest x-ray is the single most useful diagnostic tool in establishing the diagnosis of these foregut cysts. It demonstrates the cyst in the mediastinum displacing or compressing the trachea, bronchi or esophagus. There may also be evidence of hyperinflation, atelectasis or infection in either lung. Useful adjuncts include esophagography, CT scan, bronchoscopy, and echocardiography.

Treatment of foregut cysts is always complete surgical excision, traditionally through a thoracotomy. This provides relief of symptoms and definitive treatment. If the entire cyst is excised, there exists no risk of recurrence. If part of the epithelial lining is left behind, these children are at risk for recurrence and must be monitored with serial films.

**Tracheomalacia**

Tracheomalacia is generalized or local structural anomaly of the tracheal wall with deficiency or weakness of its cartilaginous framework and widening and dysplasia of tracheal membrane resulting in excessive narrowing of the trachea during expiration or in any increase intrathoracic pressure. There are following synonyms of tracheomalacia found in the literature: expiratory tracheal stenosis, tracheal dyskinesia, expiratory prolapse, collapse, expiratory invagination.

Morphological basis of the anomaly is the defect of elastic fibers and smooth muscle in the walls of the trachea and bronchi, shortening and degeneration of cartilage rings, widening and dysplasia of membranous wall. Cartilage rings were often incomplete, cartilage to muscle ratio was reduced, and there was an increase in the length of the membranous portion of the trachea which is the weakest part of the tracheal wall. In severe cases, the normal cartilage-to-membranous wall ratio of 4–5:1 can decrease to 2:1. These structural features cause the instability of the trachea during the respiration, especially forced breathing and cough. At exhalation the intrathoracic pressure increases and tracheal wall prolapses into the lumen that results in its narrowing down to a complete collapse. Narrowing of the cervical segment of trachea occurs, on the contrary, during forced inhalation. Besides the prolapse of walls of the trachea there is also a convergence of the free ends of cartilage rings that enhances stenosis. This "functional" tracheal stenosis leads to disruption of ventilation and impedes normal expectoration (discharge of secretions). The result is respiratory failure and joining of airway and lung infections.

There are two distinct forms of tracheomalacia – primary (idiopathic and associated with esophageal atresia) and secondary (due to compression or infection). Secondary TM can be congenital and acquired. The disease also is classified into diffuse and localized (up to 1/3 of tracheal length) forms. The morphological signs of tracheomalacia are found in almost all infants operated on esophageal atresia with tracheoesophageal fistula, and 25% of them may require surgical correction. Secondary tracheomalacia may be acquired or possibly associated with another congenital anomaly which compresses the airway wall during its development or growth. A localized area of a soft tracheal cartilaginous wall which develops because of pressure from a vascular ring or a parabronchial tumor or cyst may cause symptoms after the primary lesion is diminished. In infants and children, secondary tracheomalacia may present during the treatment of acquired or congenital severe pulmonary disease such as bronchopulmonary dysplasia, recurrent bronchitis and cystic fibrosis. Tracheomalacia is classified into three degrees by narrowing of the lumen: I - ½, II – ½ -2/3, III - from 2/3 to complete obstruction.

Clinical symptoms of tracheomalacia depend on the location, the length of the abnormal airway segment and the severity of the structural abnormality. Stridor is often insidious during the first few weeks of life. In 60% of the cases, symptoms appear before the age of 3 months, whereas in the remaining cases, the symptoms appear by 1 year of age. The hallmarks of this condition are: prolonged expiratory phase with wheezing, harsh barking cough, attacks of cyanosis, apneic spells and recurrent airway infections. Another characteristic of this dynamic condition is variability in both the symptoms and their intensity. Cyanosis and apneic spells often occur during feeding, coughing and crying. They are typically interspersed with normal periods of quiet sleep. The apneic spells are usually the most prominent clinical features of the disease because of their life-threatening nature. Severe obstruction is accompanied by a forced position as neck hyperextention leading to ease of breathing due to straighten and a certain stabilization of the trachea. Stridor occurs if obstruction is 2/3 of lumen and more, can be a non regular and increases when respiratory infection joins. Inability to extubate may be the symptom of severe tracheomalacia in infants operated on for esophageal atresia. These babies can easily breath through the tracheal tube but any attempt of extubation lead to severe respiratory failure. Endotracheal tube in such cases acts as a tracheal stent. More typically, infants operated on for esophageal atresia correction present with respiratory symptoms in 2 - 3 months after operation when their physical activity increases. Later disability caused by morphological changes in the respiratory system (pulmonary emphysema, recurrent pneumonia).

For the diagnostic approach, simple imaging studies, beginning with anteroposterior and lateral chest X-rays, and an esophagram using water-soluble contrast medium may be performed. Chest X-ray films can reveal a variety of anomalies, such as pneumonia, atelectasis from mucostasis or obstructing emphysema. In elder children lateral fluoroscopic view of the trachea is helpful to assess the caliber of the air-filled trachea at the different phases of respiration. Esophagography will rule out recurrent tracheo-esophageal fistula, anastomotic stricture, gastro-esophageal reflux or compression stenosis of the trachea due to vascular rings. Bronchoscopy is the most accurate means to evaluate a patient with suspected tracheomalacia. During bronchoscopy, the infant must be breathing spontaneously so that the effect of respiratory phases and coughing can be properly assessed. Marked collapse of the trachea is seen only on forced expiration or during coughing (Fig. 8). Cine CT-ultrafast is a new method of imaging which was recently introduced by the Imatron L-100 scanner which can image up to eighth level in 0.224 seconds and repeat these scans at short time intervals. The potential for acquiring serial 0.05 s images of the trachea is unique to the ultrafast CT and facilitates the identification of functional abnormalities of the trachea in infants and children.

Differential diagnosis is performed with infectious and allergic respiratory diseases (recurrent obstructive bronchitis, recurrent pneumonia, asthma). It should be remembered that these diseases may "mask" tracheomalacia. Because wheezing is a common symptom in these patients, many are mistakenly treated for asthma for long periods. It is now believed that the incidence of asthma is not higher in patients with tracheomalacia than in the rest of the population, so this diagnosis should always be reconsidered in these patients when they show no convincing response to an adequate trial of bronchodilators. In children after correction of esophageal atresia tracheomalacia can be combined with gastroesophageal reflux and recurrent fistula.

Treatment of tracheomalacia depends in large part on the severity of the disease. In children with a minor degree of collapse and mild or moderate symptoms, surgery is not necessary. Usually by 1 year of age, mild respiratory symptoms will resolve because of an increase in the size and rigidity of the trachea. Medical management is directed to the treatment of respiratory infection. Respiratory physiotherapy, bronchodilators and in-hospital observation may be useful.

A surgical approach is required for patients with severe tracheomalacia. Indications for surgery are stridor, tracheal collapse of 2/3 or more, apnea (even a single episode), recurrent pneumonia and bronchitis, inability to extubate the trachea. Choice of operations defined by mechanism of airway obstruction. The narrowing of the tracheal lumen mostly due to prolapse of anterior wall is the indicatuion for aortopexy. Aortopexy has proved to be a safe, expedient method of treating tracheomalacia in most patients with severe tracheomalacia. The aim of aortopexy is to change the cross-sectional profile of the trachea from an elipse to a circle so that its walls will not appose during ventilation and coughing, and at the same time move the trachea away from the esophagus so that when full it cannot compress the airway. The suspension of the aorta vascular structure from the under-surface of the sternum is done without disrupting the fascia between it and the adjacent trachea which acts as a suspension ligament to change the configuration of the trachea.

If the leading factor of expiratory stenosis is the prolapse of tracheal membrane, tracheoplasty is performed (reinforcement of posterior tracheal wall by means of grafts of different materials: autopericardial, fascial or artificial patch).

The combination of both factors of airway obstruction and a significant degree of stenosis requiring a combination of aortopexy and tracheoplasty.

Prognosis of surgical treatment of tracheomalacia at the correct indications and mode of operation is favorable, positive results achieved in about 95% of cases.

**Tracheal diverticulum.** This is an extremely rare anomaly, usually arising from the right posterolateral surface of the trachea, that may give rise to symptoms only late in adult life when it becomes infected. Because of this, it has been suggested that it may be an acquired rather than a congenital lesion, but a few reports of congenital diverticula exist. Some consider this malformation as a blind-ending tracheal bud, i.e., the lesser form of an accessory tracheal bronchus.

**Esophageal atresia (EA)** is themalformations related to defective septation of the primitive foregut. This is the most common among other foregut malformations, with an incidence of 1 in 3000 to 5000 live births. In 80% of cases, the esophageal atresia is accompanied by a tracheoesophageal fistula (TEF) between the lower trachea or carina and distal esophagus.

A working classification based on the frequency of each anomaly is of the greatest practical value to the neonatal surgeon.



Classification of EA and/or TEF (from left to right):Esophageal atresia with distal tracheoesophageal fistula (82.8%); Esophageal atresia without fistula (8.4%); Esophageal atresia with proximal fistula (2.1%); Esophageal atresia with proximal and distal fistulas (3.4%); Tracheoesophageal fistula (H-type) without atresia (3.3%).

The sporadic reports of vertical and transverse familial cases of EA and TEF suggest a polygenic hereditary etiology. The best estimate of risk of recurrence for parents of a single affected child is 0.5–2.0%, rising to 20% if another sibling is born with EA. The vertical transmission risk is 3–4%. A 10% incidence of non-specific chromosomal abnormalities (translocations, deletions and duplications) has been noted. However, only trisomies 18 and 21 show any definite association with EA and TEF.

Associated anomalies are seen in over half of all newborns presenting with EA and TEF. Congenital heart disease (27%) is the commonest comorbid condition and has the greatest impact upon survival. Recently aortic arch anomalies have been shown to occur frequently in association with long-gap EA and TEF. Other common associated abnormalities include urogenital (18%), skeletal (12%), anorectal (12%), and other gastrointestinal conditions (9%), most notably duodenal atresia. Several phenotypic variants have been reported in association with EA and TEF. The first to be described was the VATER association, which is now encompassed by the VACTERL acronym. The presence of three or more of the features is essential to define the association. The CHARGE association (coloboma, heart disease, atresia choanae, retarded development, genital hypoplasia, and ear deformities with deafness), is another constellation of phenotypes associated with EA and TEF. EA and TEF are also recognized in the SCHISIS association (exomphalos, neural tube defects, cleft lip and palate, and genital hypoplasia).

**Clinical presentation and diagnosis**. A newborn with EA is often noted to have difficulty clearing saliva. Episodes of coughing, choking and even transient cyanosis may be observed shortly after birth. These signs are frequently overlooked and attempts to feed the infant result in immediate respiratory distress. The diagnosis is readily confirmed by the failure of passage of a firm nasogastric tube. A characteristic resistance is felt at the blind ending upper esophageal pouch, and the tube cannot be introduced into the stomach. A plain X-ray, which should include the chest and abdomen, demonstrates the nasogastric tube coiled in the upper pouch. An associated TEF is confirmed by the presence of gas-filled intestinal loops below the diaphragm. In isolated or pure EA, a featureless gasless abdominal X-ray is observed. The presence of a double bubble on the abdominal film suggests associated duodenal atresia. A careful search for associated abnormalities is mandatory, specifically checking for patency of the anus. The cardiovascular system should be examined to exclude a major congenital heart defect whose treatment may take priority over correction of the EA.

**Antenatal diagnosis**. Fetal diagnosis is now possible in cases of EA and TEF. The classical ultrasonographic features of EA and TEF in the fetus are absence of the stomach bubble and associated hydramnios. However, prenatal detection rates remain low (9–24%), and there is a high false-positive rate, with over half of all cases proven not to have EA after birth.

**Management.** Having established the diagnosis, i.v. fluids are started, endotracheal intubation performed and a sump suction catheter introduced into the upper pouch to allow continuous or frequent intermittent aspiration of secretions. Broad-spectrum antibiotics and Vitamin K should be administered. The baby is transferred to the neonatal surgical unit. An echocardiogram performed prior to surgery will alert the surgeon and anesthetist to an underlying cardiac defect that may adversely influence prognosis, and may influence the operative approach by identifying the side of the aortic arch. Surgery is ideally performed within the first 24 hours in an otherwise healthy newborn.

The infant is positioned for right thoracotomy in a lateral or postero-lateral position. Some surgeons choose the surgical access to be on the opposite side of artic arch. The chest is opened through the fourth interspace. The pleura is carefully separated from the ribs to commence an extra-pleural approach towards the fistula. The azygos vein is mobilized and controlled with suture slings. The distal esophagus is identified and the TEF is dissected and divided and the defect of tracheal wall is sutured. Then upper esophageal pouch is identified and mobilized to diminish the gap. An upper pouch fistula may be identified at this stage, and should be repaired. In most cases of EA with distal TEF, a primary anastomosis is possible, although occasionally considerable tension is required to complete the repair. Chest drain is placed with the tip near the anastomosis. The chest drain should be attached to under-water drainage.

Should additional surgical pathology be present, such as duodenal atresia or imperforate anus, these should be dealt with accordingly under the same anesthetic in the stable infant.

Surgical management of neonates with “pure” EA is challenging and controversial. The gap between the blind end of proximal and distal esophagus is very long in most cases making the primary anastomosis impossible. The majority of pediatric surgeons consider delayed primary anastomosis of the native esophagus, the optimum approach. The infant with isolated EA is initially managed by feeding gastrostomy and continuous suctioning of the upper pouch. After a period of approximately 3 weeks, the extent of the gap can be assessed by fluoroscopy. A distance of less than two vertebral bodies separating upper and lower pouches is ideal to attempt delayed primary anastomosis. A spiral, circular or staged myotomy or upper pouch flap may be required to achieve an anastomosis. Extra length can be obtained on the lower pouch if necessary by proceeding to laparotomy and performing either a Scharli lesser curve myotomy or a Collis gastroplasty. If a primary anastomosis is still not possible the option is esophageal replacement (esophagocoloplasty) at the age of 10-12 months.

**Congenital isolated tracheoesophageal fistula (H-fistula).** An isolated tracheoesophageal fistula is a rare malformation with prevalence about 1:100000 newborns and seen in about 5% of all esophageal malformations. American researchers D.S. Lamb and A. Fatal were the first to describe the defect as early as 1873. The symptoms are recurrent coughing and choking, especially during feedings. The diagnosis is often delayed, resulting in recurrent pneumonia. A distended abdomen with airfilled loops may also be noted. When the diagnosis is suspected, fluoroscopy is performed in the prone position with contrast injection through a nasoesophageal tube starting in the mid-esophagus and gradually moving up. Falsenegative studies may occur. Rigid bronchoscopy is essential to confirm the diagnosis and it facilitates the operation. The fistula, usually located in the lower cervical area, is cannulated during bronchoscopy. Diagnostic studies (esophagography, fistula cannulation followed by chest X-ray, bronchscopic picture) are indicated in figure to facilitate intraoperative identification and minimize the amount of dissection, thereby decreasing the risk of recurrent laryngeal nerve injury. Simple ligation and division of the fistula through a cervical approach give excellent results with few long-term complications. Attempts at endoscopic occlusion of the fistula with glue, laser, or other means is associated with a high recurrence rate and do not appear warranted in H-type fistulas that can be divided through a cervical approach. These techniques may have their place in the management of recurrent TEF. About 10% of H-fistulas are too low to be divided through a cervical approach and require thoracotomy or thoracoscopy.

**Congenital malformations of the lungs**

Congenital malformations of the lung are uncommon but extraordinarily diverse in their presentation and important to all physicians and surgeons who care for infants and children. The spectrum of presentation includes antenatal diagnosis of malformations to presentation in adulthood and ranges from asymptomatic to immediately life threatening.

Formation of the bronchopulmonary system begins at 3 - 4 weeks of gestation from the caudal foregut segment, i.e. from one tube, which is then delimited on the trachea and esophagus. Not by chance, some authors consider the malformations of the esophagus (atresia, fistulas) as defects of the respiratory system. On 7 – 8th month of gestation in the process of encoded intensive development of embryo alveolar ducts give rise of acini, and capillaries deepen under an epithelium forming alveoli with aero-hematic barrier as a gas exchange organ.

Respiratory and vascular components of lungs complete their development, mainly before 26 weeks of gestation, but for the valuable functioning and postnatal life-support 30 weeks is needed.

After birth during 1 - 4 months an air-vascular organ continues its development, the number of alveoli increases as well as the lumen of arterioles, blood pressure decreases (since 10th day) with the reduction of fetal circulation. Lungs as an organ complete their formation by 7 - 8 years and continue its growth to 25 - 27 years.

Malformations of the respiratory system occur for unknown reasons. However, endogenous and exogenous teratogenic factors are distinguished among them. First include heredity, chromosome abnormalities, endocrine diseases, biological inferiority of the germ cells, including the pregnancy in late reproductive period of mother and father in old age and the last involve physical (mechanical, electromagnetic, radiation), chemical (any poisons, hormonal, defects of feed), biological (bacteria, viruses).

**Classification**. The first classification of malformations of lung was developed by Schneider and Schwalbe in 1912, which distinguished between three types of underdevelopment of the lung tissue: agenesis (complete absence of one or both lungs), aplasia (absence of the lung with the presence of rudimentary main bronchus) and hypoplasia (abortive development of one or two lungs). In Ukraine the classification of А.М. Sazonov et al (1981) is rather popular, which, in particular, underlines vascular anomalies in the spectrum of defects of the respiratory system. This classification is as follows.

First group consists of anomalies caused by combined disorders of several structures: agenesis, aplasia, hypoplasia of lungs or lobe (simple and cystic, including polycystic lung) and additional lung lobes.

Second group includes anomalies caused by predominant violation of broncho-epithelial branching: tracheobronchomegaly and tracheobronchomalacia; stenosis of the trachea and bronchi; lobar emphysema; diverticula of trachea and bronchi; broncho-esophageal fistula; bronchogenic cyst; congenital bronchiectasis; hamartohondromas.

Third group includes anomalies of the pulmonary vessels: aplasia and hypoplasia of vessels; aneurysms and arteriovenous fistulas; anomalies of pulmonary venous return.

Fourth group involves anomalies of systemic vessels: pulmonary sequestration (extrapulmonary, intrapulmonary); abnormalities of bronchial vessels (aplasia, hypoplasia) and azygos vein lobe.

And, finally, the fifths group comprises anomalies of other tissues and organs : dermoid cysts; teratomas; mucoviscidosis.

**Pulmonary aplasia and agenesis**

Pulmonary agenesis and aplasia are rare malformations. Their incidence has been estimated to be 1 in 10 000–15000 autopsies. The malformation have been known since 1762, when the Italian anatomist Morgagni first described it. Since then, about 300 cases have been mentioned in the literature with 2–4 cases per report. Lung agenesis and lung aplasia should be viewed as separate malformations. The mortality of children with lung aplasia is 33% during the first year of life and 50% in the first 5 years of life. Lethal outcomes are usually the result of infection of the single lung and concomitant severe heart and vessel malformations. The course of pulmonary agenesis is more favorable.

**Lung aplasia** is described as an absence of pulmonary parenchyma and pulmonary artery but with a rudimentary main bronchus and a well-formed tracheal bifurcation. If present, the rudimentary pulmonary parenchyma is hypoplastic and atelectic.

**Relevant anatomy and pathophysiology of lung aplasia.** The presence of the bifurcation and carina is essential for the pathogenesis of lung aplasia as these provide fixation of the trachea to the pericardium and aortal arch, which results in a shift of the trachea following the heart and great vessels. Tracheal kinking right above the bifurcation and at the upper thoracic aperture narrows the trachea even further. Such a tracheal shift and double kinking does not happen in lung agenesis where the tracheal bifurcation and its ligaments are absent. The superior vena cava is kinked as well. Tracheal kinking and narrowing in the presence of the main bronchus stump (which may be rather long up to 3 cm) transforms the laminar airflow into a turbulent flow. It also increases resistance and dead space and promotes the piling up of infected bronchial secretions. Normal diaphragmatic movements promote a mediastinal shift into the negative pressured empty pleural cavity with lung herniation and emphysema of herniated segments or lobe due to kinking of the respective bronchi. The perfusion in herniated segments is almost absent. Together with the pressure of the brachiocephalic vessels on the upper trachea, such herniation results in a further worsening of tracheal compression and obstruction. The hemodynamic changes include kinking of the superior vena cava and innominate vein resulting in a decrease of brain venous outflow. Left-to-right blood shunting due to an opened foramen ovale or patent ductus arteriosus results in hypervolemia of the single lung. Such changes cause early and progressive respiratory distress with infection of the single lung in children with lung aplasia.

**Clinical presentation and diagnosis.** The signs of respiratory failure develop together with the increase in physical activity by the 3rd month of life or after respiratory infections. The anatomical peculiarities of the malformation expedite such complications. If the patients overcome these complications in their early years, respiratory distress develops at the age of 3–4 years and progresses, limiting normal physical activity, especially during the cold season. The main symptoms are shortness of breath, coughing, wheezing in the lungs, noisy breathing, low physical exercise tolerance, frequent respiratory infections, delay in physical growth, headaches, acrocyanosis, chest pains and palpitations. Most of the manifest or enhanced with increased physical activity of children in the first 2 - 3 months of life.

On physical examination one can reveal shift of the heart to affected side, deformity, asymmetry, retraction of the chest on the side of the missing lung.

Plain chest radiograph shows mediastinal shift to the side of the lesion, the affected hemithorax is opaque, while the opposite hemithorax is of increased transparency and enlarged. Lateral X-ray shows the anterior mediastinum enlightenment due to prolapse of single lung. Verification of the diagnosis requires the use of cardioangiopulmonography, CT and MRI. Digital subtraction angiography reliably establish the absence of lung, agenesis of the pulmonary artery, kinking of venous trunks, the position of the aorta and major vessels in relation to the airway

Trachoebronchscopy shows rotation and anterior vascular compression of the trachea. Tracheobronchography is risky because of the threat of enhance of respiratory failure.

Differential diagnosis should be made with pulmonary agenesis, diaphragmatic hernia, atelectasis of the lung, congenital emphysema, chylothorax and pleuritis, bronchial obstruction due to foreign body.

Treatment. For many years it was believed that patients with lung aplasia subject only conservative treatment of respiratory infections and observation. Recent studies point to the possibility and the need for surgical treatment of respiratory disorders in these patients (C. Dohlemann at al, 1990, F. Becmeur et al, 1995, D. Krivchenya and singing., 1995, 2000, 2007). Considering that the major pathogenetic factors of respiratory distress in the lung aplasia are kinks and compression of airway, displacement, rotation and kinking of great vessels and heart, instability of mediastinum and emphysema of single lung, the following surgical techniques offered:

- cephalad diaphragmatic cupola translocation on the affected side. The operation results in a decrease of the affected hemithorax volume and of the ipsilateral diaphragmatic cupola function by approximately 30%, with normalization of the mediastinal position and mediastinal stabilization. This prevents kinking of the tracheal and large vessels and vascular airway compression. It helps to stop the progression of emphysema in the single lung and promotes tolerance of physical exercise. The postop chest X-ray and diagram is shown on Fig. 14b.

- anterior aortopexy to relieve airway compression caused by displacement of cardiovascular complex;

- combination of these operations accompanied by possible resection (amputation) stump bronchus

**Pulmonary agenesis.** The carina, bronchi, pulmonary vessels and parenchyma are absent in this malformation. In lung agenesis, the single main bronchus is a direct continuation of the trachea. The tracheal bifurcation, the carina and the contralateral main bronchus are all absent. The trachea and main bronchus are less closely connected with the large vessels, which is why their shift to the affected side is less marked, and therefore there is no tracheal and bronchial kinking and stenosis. However, there is a possibility of tracheal compression by the large vessels, especially in the case of right lung agenesis.

Symptoms may appear from the first years of life as a susceptibility to respiratory infections. The chest is asymmetric with diminution of affected hemithorax. The mild-symptom course is possible, and sometimes the malformation is a random finding. Patients presenting late usually have readily detectable flattening and reduced movement of the chest wall on the affected side, with reduced air entry on auscultation, although this often sounds surprisingly better than expected. There may be some breathlessness on exertion and the chest wall deformity may be quite pronounced, with an associated secondary scoliosis.

Imaging techniques combined with rigid bronchoscopy allow a precise diagnosis to be made. On the chest radiograph, there is considerable mediastinal shift and the involved hemithorax is small, with narrowed intercostal spaces. Absence of the ipsilateral mainstem bronchus or the pulmonary artery are definitive diagnostic findings and this can be established by endoscopy, angiography, ECHO or axial imaging techniques. A healthy lung is of increased transparency, enlarged and hyperinfated, thus may simulate congenital emphysema (risk of erroneous operation on a single lung!).

Management is initially limited to supportive treatment (including oxygen if necessary), correcting associated malformations, and the prevention and treatment of respiratory infections. In some cases tracheal stenosis or vascular compression may coexist and require treatment. Some patients with severe respiratory distress due to mediastinal shift, tracheal kinking and its vascular compression may require surgery. Cephalad diaphragmatic cupola translocation on affected side and anterior aortopexy may be considered as effective procedures.

**Pulmonary hypoplasia** is defined as an underdevelopment of all structural elements of the lung (bronchi, vessels and parenchyma). The malformation is believed to be caused by derangement of development of primary lung buds.

Pulmonary hypoplasia is almost always accompanied by hypoplasia of the corresponding pulmonary vessel. This follows obviously from the second rule of lung embryogenesis, in which the vasculature follows the bronchial development.

Hypoplasia as an isolated phenomenon is rare. More commonly, pulmonary hypoplasia is associated with conditions that interfere with lung growth including following: conditions leading to an egress of lung fluid (severe oligohydramnios, compression of the thoracic cage and abdominal contents by the uterus, laryngotracheoesophageal cleft and large tracheoesophageal fistula); space-occupying lesions (congenital diaphragmatic hernia, eventration, lung malformations, thoracic tumors, pleural effusion, chylothorax, abdominal conditions pressing on the diaphragm); thoracic cage anomalies (Jeune syndrome, achondroplasia, scoliosis); conditions preventing normal fetal breathing movements (anencephaly, phrenic nerve agenesis).

Pathogenesis of pulmonary hypoplasia involves derangement of bronchial patency, which sustains hypoxia of the lung tissue and desolation of pulmonary vessels, pneumosclerosis and of obstructive emphysema.

Hypoplasia of the lungs may be combined with other defects, such as diaphragmatic hernia, malformations of the skeletal system, urinary tract, heart, central nervous system and the digestive system.

There are two major types of lung hypoplasia distinguished: simple and cystic hypoplasia.

Clinically, patients with pulmonary hypoplasia may present in early infancy with respiratory distress ranging from mild to severe, depending on the degree of hypoplasia. Commonly, it is the associated anomalies that draw the attention. In severe bilateral hypoplasia the thoracic cage is obviously reduced in size and characteristically bell shaped, with the base of the chest widening at diaphragmatic level to a normalsized abdomen. The patient is tachypneic, with restricted chest wall movement, and in respiratory distress. Less severe degrees of hypoplasia—unilateral or bilateral—may present later with persistent tachypnea or disproportionate shortness of breath with exercise.

Occasionally, the abnormality is coincidentally noticed on examination or chest radiograph when the patient presents with an intercurrent infection. In unilateral hypoplasia, the chest cage appears asymmetric, with diminished air entry and chest expansion on the side of the lesion together with mediastinal shift to that side. The chest radiograph confirms mediastinal deviation and a lung that often appears hyperlucent. Isotope scanning usually reveals a greater impairment of perfusion than ventilation on the side of the lesion.

The diagnosis and treatment of pulmonary hypoplasia is dependent on the underlying etiology. For example, severe respiratory impairment can be seen with pulmonary hypoplasia from a CDH. In these cases, stabilization of the underlying physiologic impairment is the priority.

Treatment is initially directed at any associated anomalies, with particular focus on congenital cardiac lesions. If an operation is needed to remove a nonfunctioning lobe or lung, every effort is made to preserve functional parenchyma.

The absolute indication for surgical treatment is local and mosaic forms of lesion with involving up to 12 lung segments with progressive and recurrent type of inflammatory course.

The method of surgical treatment depends on the amount of affected lung. Typical anatomical resection of the affected lung zones is the operation of choice for the lesion localized within one or two lobes or whole lung.

Segmental resection of the lung is indicated in the cases of confirmed cavities, both within the bronchus and parenchyma of the segment.

Sparing (atypical) resection of the affected area is indicated for local and mosaic lesion.

Combined resection are advisable to carry out in patients who have cavitary lesions in adjacent areas of the lungs.

At sparing and combined operations pneumatization of remaining lung tissue plays a positive role in the prevention of bronchial kinking and fulfilling the remaining volume of the chest cavity.

In the absence of associated lesions, unilateral pulmonary hypoplasia is compatible with normal growth, development, and survival. Long-term complications include reduced exercise tolerance, recurrent infections and sometimes a worsening chest deformity with scoliosis.

Congenital emphysema is a rare malformation that is characterized by hypertrophy, hyperinflation and overdistention of the lung parenchyma. Although usually described as affecting lobes it may affect individual segments causing an increase in parenchymal lung volume and a decrease in blood flow and potentially compromising ventilation. It may present early with respiratory distress in infants and hence require urgent surgery. Although “congenital lobar emphysema” is the commonest phrase used in the literature, it does not necessarily correspond with the anatomic location of the affected zone, especially if the superior division (S1, S2, S3) bronchopulmonary segments of the left lung are affected.

The following causes have been described as a cause of emphysema: dysplasia and lack of bronchial cartilage (bronchomalacia); folds of mucous membrane creating a ball-valve-type effect; obstruction by mucus plug; bronchial stenosis; bronchial atresia; external compression of the bronchus caused by abnormal vessels; enlarged lymph nodes. A polyalveolar lobe has also been found in association with some cases of congenital emphysema. Nevertheless it has been suggested that in up to half of all cases the exact cause of congenital emphysema remains unknown.

The diagnosis of congenital lobar or segmental emphysema is based on clinical findings, chest radiography, digital subtraction pulmonary angiography (DSA) and CT with intravenous contrast enhancement.

The differential diagnosis for such cases included pneumonia; respiratory tract foreign bodies; bronchial obstruction by aberrant blood vessels; pulmonary agenesis, aplasia, hypoplasia and dysplasia; and agenesis of pulmonary artery and its branches. Bronchoscopy was used in some cases to exclude lung aplasia or foreign bodies. Bronchography is considered dangerous particularly in those patients with an acute presentation.

The main radiographic signs consistent with congenital emphysema are as follows: a) increase of the lung transparency on the affected side with decreased vascular markings; b) shift of the mediastinum to the contralateral side (mediastinal herniation); c) pseudodextrocardia in left-sided cases; d) ipsilateral flattening of the diaphragm cupola; e) ipsilateral expansion of intercostal spaces and g) reduction of contralateral lung field with a reduction in transparency (Fig. 16)

Three types of clinical presentations were defined: decompensated, sub-compensated, and compensated. Thus typical symptoms in the decompensated form were acute respiratory insufficiency with shortness of breath at rest, skin pallor, and in critical cases, cyanosis, asphyxia and convulsions. In the sub-compensated form children had symptoms of shortness of breath, coughing, and sweating on participation in minor physical activity.

The absence of blood perfusion in the affected segments was well shown by DSA and particularly the characteristic sparing of the lingual (S4 and S5) segments in left-sided disease. Fig. 17 illustrates the typical vascular pattern of affected lung parenchyma.

Surgery is performed through a lateral fourth space thoracotomy. Lung-sparing surgery removing only abnormal lung parenchyma is the aim and implies segmental resection in most left-sided cases. Division of a patent ductus arteriosus or short ligamentum arteriosum is performed where necessary. Preservation of lingual (S4 and S5) segments of the left lung normalizes position of the thoracic organs, prevents over-distention of remaining lung parenchyma and prevents the development of a large residual postoperative pleural cavity.

Lobectomy is used for cases with whole lobe impairment that is more characteristic for right-sided lesion.

**Congenital lung cysts**

Congenital lung cysts are intrapulmonary cavities filled with fluid or air, inside lined with epithelium. Some of these may actually be intrapulmonary bronchogenic cysts or unilocular variants of congenital pulmonary airway malformations (CPAM/CCAM). Congenital lung cysts, in contrast to bronchgenic cysts, arise from more distal airways, including the alveoli, or they may be pleural in origin. The incidence of congenital lung cysts ranged from 3.5 to 5.5% among children with chronic nonspecific lung diseases.

The solitary and multiple cysts are distinguished.

The main clinical symptoms of uncomplicated lung cysts are following: coughing especially in wet cold weather, shortness of breath, frequent recurrences of respiratory infections (bronchitis, obstructive bronchitis, pneumonia), and reduced tolerance to physical activity.

Complications of lung cysts are tension, suppuration and a breakthrough into the pleural cavity. Complicated cases present with acute symptoms including respiratory distress, signs if pulmonary inflammation and suppuration, chest pain, shortness of breath, increase of body temperature, discharge of purulent sputum as "full mouth."

The diagnosis of lung cysts is made by using general clinical and instrumental examination: chest radiography, conventional tomography, CT.

The X-ray signs of cysts are: thin-walled round zone of transparency or opacification (depending on content) omitted by pulmonary vascular pattern. Tension lung cyst causes mediastinal a shift in the opposite direction. Cyst complicated by suppuration that drained into the bronchi, has a fluid level inside. Congenital solitary cysts have one cavity, otherwise multiple lungs cysts consist of many cavities.

Any localized cystic lung lesion that appears congenital and is confirmed on CT scan or MRI should be excised because of the potential risks of infection, erosion into an adjacent bronchus, pneumothorax and the inability to distinguish it from the type I pleuropulmonary blastoma.

Operations are carried out regardless of the age of the child in compliance with the principle of organ preservation and maximum functional lung tissue.

**Congenital Pulmonary Airway Malformations (congenital cystic adenomatoid malformation)**

Congenital pulmonary airway malformations (CPAM), previously termed congenital cystic adenomatoid malformations (CCAM), considered a hamartomatous lesion of the bronchial tree by some, whereas others favor a localized arrest in the development of the fetal bronchial tree as the etiology. The incidence of CCAM/CPAM has been estimated at 1 per 25,000 to 35,000 or even 100,000pregnancies. While relatively uncommon, CPAM constitute 10–30% of congenital lung malformations in most series, with a slight male predominance. Histologically, CPAM are composed of disorganized cysts lined with ciliated cuboidal or columnar epithelium. Involvement is generally unilobar with a slight predilection for the lower lobes, with right and left sides affected equally. Typically, they have normal pulmonary arterial and venous blood supply and communicate with the tracheobronchial tree.

**Clinical Features.** In the antenatal and immediate postnatal period, the predominant physiologic consequences of a CPAM result from compression of the mediastinum and adjacent normal lung by the mass lesion. Because of the mass effect, polyhydraminos, mediastinal shift, pleural effusions, and fetal hydrops can be identified in the antenatal period. In contrast, the mortality approaches zero for a fetus with a CPAM but without hydrops. In addition, spontaneous resolution in utero of an antenatally diagnosed CPAM is reported to occur in up to 15% of patients. Following delivery, the major risk to the newborn with a CPAM is respiratory distress as a consequence of compression of the adjacent normal lung and mediastinum by the CPAM. Far more commonly, infants born with a CPAM either remain asymptomatic or have persistent mildly increased work in breathing that interferes with feeding.

**Diagnosis**. *Prenatal diagnosis* by sonography is relatively common, which demonstrates an echogenic pulmonary mass with displacement of adjacent structures. Using antenatal sonography, CPAM is defined as either macrocystic (greater than 5-mm diameter cysts) or microcystic (solid or less than 5-mm diameter cysts), but the natural history of CPAM is determined more by overall size and degree of compression of adjacent structure than by their gross appearance. The prenatal differential diagnosis includes congenital diaphragmatic hernia, pulmonary sequestration, and bronchogenic cyst. Sonographic findings associated with CPAM include polyhydramnios, mediastinal shift, pleural effusions, and fetal hydrops. *Postnatal diagnosis* can often be made by plain chest radiographs; however, in a stable patient, CT scans are often helpful to define anatomy and particularly to identify aberrant systemic blood supply that is more suggestive of a pulmonary sequestration. A CT scan is more accurate than plain chest radiography in confirming complete resolution of an antenatally diagnosed congenital pulmonary lesion that is not seen on postnatal plain chest radiography (Fig. 19).

**Management**. *Fetal therapy.* For congenital cystic pulmonary lesions, it is clear from the literature that provided the fetus does not experience hydrops or physiologic distress, treatment should be expectant management with term delivery and postnatal evaluation. Administration maternal steroids to any affected fetus less than the estimated gestational age of 34 weeks to help fetal lung maturation is reasonable.In fetuses with ultrasonographic evidence of deterioration due to fetal hydrops, intervention should be considered. If the fetus is in the second trimester, several options exist for fetal intervention. Ultrasound-guided intrauterine thoracoamniotic shunting for a macrocystic CPAM with a large cyst has the best outcome with the lowest fetal and maternal risk. If hydrops develops during the third trimester, an EXIT procedure with thoracotomy and lobectomy using placental bypass can permit safe resection and avoid respiratory collapse.

*Postnatal therapy.* Prompt surgical resection should be performed in any symptomatic newborn. In those patients who remain asymptomatic in the newborn period, delaying resection until infancy is reasonable and allows somatic growth that may facilitate the ease of pulmonary resection. Asymptomatic but persistent CPAM should be resected during infancy to prevent complications of recurrent infections or malignant degeneration. Resection often includes formal lobectomy; however, for small CPAM, nonanatomical resection is reasonable. Traditionally, resection was performed through open thoracotomy but recent reports have demonstrated excellent outcomes for minimally invasive pulmonary resection.

**Bronchopulmonary Sequestrations**

Bronchopulmonary sequestrations are defined as either an intrathoracic or subdiaphragmatic mass of nonfunctional pulmonary tissue that lacks communication with the tracheo-bronchial tree of the normal lung. Histologically, pulmonary sequestrations demonstrate immature lung development, often resembling more peripheral lung parenchyma. Intrathoracic sequestrations are further categorized as either extralobar or intralobar, depending on whether the lesion is invested by its own pleura or that of the adjacent normal lobe, respectively. The arterial blood supply is anomalous, most commonly arising from the descending thoracic aorta. Venous drainage for intralobar sequestrations is most commonly via the pulmonary veins, whereas venous drainage for extralobar sequestrations is typically systemic via either the azygous or hemiazygous veins, inferior vena cava, or directly into the atrium.

**Clinical features.** Both intralobar and extralobar sequestrations can present in the newborn period with either respiratory distress due to mass effect or congestive heart failure because of arteriovenous shunting within the sequestration. Most intralobar sequestrations present later in childhood due to recurrent pulmonary infections from inadequate tracheobronchial drainage, or less commonly with hemorrhage. The abnormally developed lung tissue is ineffective in gas exchange and therefore is not of benefit to the patient.

**Management** of pulmonary sequestrations is surgical.The goal of surgical resection is to remove only the abnormal portion of lung with obvious division of the aberrant artery supplying the sequestration. Care should be given to the division of this artery because it is big in diameter, originates directly from thoracic aorta, has high (the same with aorta) blood pressure inside and may contract after division hiding within the mediastinum and causing uncontrolled arterial bleeding been inadequately ligated. For intralobar sequestration, particularly if previously infected, this often necessitates lobar resection.

**Congenital Diaphragmatic Hernia**

Congenital diaphragmatic hernia (CDH) is a developmental abnormality of the diaphragm that allows the abdominal viscera to enter the thoracic cavity. It occurs with an overall incidence ranging from 1 in 2000 to 4000 live births. There is a slight male preponderance with a ratio of 1.5 : 1. Most CDHs occur through a posterolateral defect (Bochdalek hernia), with 90% of these on the left side, 10% on the right and less than 1% bilateral. Less than 5% of CDHs are located anteriorly (Morgagni hernia). A number of syndromes are associated with CDH. In some syndromes, such as Fryns syndrome and Donnai-Barrow syndrome, CDH is present in a high percentage of affected individuals. Others include Beckwith-Wiedemanch, Simpson-Golabi-Behmel, Coffin-Siris, and Denys-Drash syndromes. CDH has also been reported with trisomies 9, 13, 18, 21, and 22.

Embryogenesis of CDH has been described as a failure of the closure of the pleuroperitoneal canal, which occurs during gestational week 8. Consequently, the abdominal viscera herniates into the thorax, which is thought to cause pulmonary hypoplasia by compression of the growing lung. Furthermore, a toxicological nitrofen model of CDH has shown that abnormalities in the contralateral lung as well as the ipsilateral side are present even before the diaphragm starts to develop.

The *prenatal diagnosis* of CDH has been greatly facilitated by the improved technology and interpretation of prenatal imaging studies. CDH is readily identified on routine prenatal ultrasonography as early as 18 weeks of gestational age. Displacement of the mediastinum, the absence of a stomach bubble in the abdomen, and the presence of abdominal organs in the chest are the signs of fetal diaphragmatic hernia. A right-sided CDH is more difficult to identify because the echogenicity of the fetal liver is similar to that of the lung. Identifying the gallbladder in the fetal chest may be the most reliable sign in these cases. Prenatal magnetic resonance imaging has been shown to be effective in confirming the diagnosis of CDH and detecting additional information that may affect prognosis. The two most commonly used prenatal predictors of postnatal morbidity in infants with CDH are liver position and lung-to-head ratio (LHR). An intrathoracic location of the liver and a low LHR (<1.0) predicts a poor prognosis.

*Postnatal presentation and diagnosis*. The onset and severity of symptom depends on the amount of abdominal viscera in the chest and the degree of pulmonary hypoplasia. The most severely affected infants present with respiratory distress at birth. Other infants with CDH develop cyanosis, tachypnea, and grunting respirations within minutes or hours after birth. Physical examination reveals a scaphoid abdomen, an increased anteroposterior diameter of the thorax, and mediastinal shift. Breath sounds are absent on the affected side. The definitive diagnosis of CDH is made postnatally by plain radiography of the chest and abdomen by demonstration of air-filled loops of the bowel in the chest and a paucity of gas in the abdomen (Figs. 20, 21). The diaphragmatic margin is absent, there is a mediastinal shift to the opposite side, and only a small portion of lung may be seen on the ipsilateral side.

Although most CDH infants present in the first 24 h of life, 10–20% of the affected infants present later. The symptoms and signs of those patients are nonspecific and include recurrent chest infections, vomiting, abdominal pain, diarrhea, anorexia, failure to thrive, or an abdominal chest X-ray in an asymptomatic patient. Some children present acutely with volvulus or strangulation or acute respiratory distress. Chest X-ray with an in situ nasogastric tube is reliable for the diagnosis.

Differential diagnosis includes cystic lung disease and mediastinal cystic lesions (e.g. cystic teratoma, neuroenteric, bronchogenic, and thymic cysts). The diagnosis of CDH can also be confused with other congenital thoracic conditions, such as eventration of the diaphragm, anterior diaphragmatic hernia of Morgagni, congenital esophageal hiatal hernia, and primary agenesis of the lung.

**Management**. *Preoperative management.* An infant with respiratory distress requires endotracheal ventilatory support. Bag-and-mask ventilation should be avoided to minimize gaseous distention of the stomach and intestines, which would further compromise lung function. Prompt endotracheal intubation and limitation of ventilation pressures are essential. The concept of “gentle” ventilation and permissive hypercapnia was proposed to minimize barotraumas by strictly limiting the peak inflation pressure. A nasogastric tube is passed and placed on suction and a chest radiograph is done. The basic tenets of management for the infant with CDH are based on the support of the cardiorespiratory system. First, the patient is placed on monitoring equipment, intravenous access established and pre- and postductal oxygen saturations recorded with pulse oximetry. Postductal saturations provide caregivers with an indication of the severity of pulmonary hypertension and right-to-left shunting. High-frequency oscillatory ventilation (HFOV) is a mode of ventilation that can be effectively used in CDH infants to maintain preductal oxygenation while avoiding hyperventilation and barotraumas. Infants who fail to respond to optimal therapy may be placed on extracorporal membrane oxygenation (ECMO). Inotropic support to improve the circulatory status of the patient may also be considered for patients with low mean arterial pressure and derangements in systemic perfusion. Excessive amounts of intravenous fluid should also be avoided because pulmonary edema with worsening pulmonary hypertension and gas exchange may ensue.

*Surgical correction*. The philosophy regarding the timing of surgical repair has generally shifted from emergent repair to a delayed approach after stabilization of the infant. Although the abdominal approach is widespread among surgeons as it offers good exposure, easy reduction of the abdominal viscera, and recognition and correction of associated gastrointestinal anomalies, we have recently proposed approach through thoracotomy in both right and left-sided CDH. This is based on the concept of thoracalization of abdominal cavity with patch diaphragmatic defect repair. This allows keeping dome-shaped diaphragm, preventing the increase of intraabdominal pressure and decreasing the big volume of “empty” hemithorax on affected side thus protecting the hypoplastic lung from hyperinflation and barotrauma.

Small diaphragmatic defects can be sutured by direct sutures of the edges of the defect after the contents of hernia are gently reduced in the abdomen.

If the defect is large, it may not be possible to repair it using direct sutures. Various techniques have been described and include the use of prerenal fascia, rib structures, the lastissimus 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 have been advocated for repair of large defects. The most commonly used prosthetic material presently are surgisis soft tissue graft, PTFE-graft.

*Postoperative care.* Ventilatory support should be continued postoperatively. The vital signs are monitored closely with regular blood gas analyses and monitoring of preductal and postductal oxygenation. Some infants show improvement in oxygenation in the immediate postoperative period, the so-called honeymoon period, but deteriorate 6–24 h later. This deterioration is due to pulmonary hypertension and persistent fetal circulation with an increase in pulmonary artery resistance, elevated pulmonary artery pressure, and right-to-left ductal and preductal shunting leading to hypoxemia.

Despite advances in care, intervention, and technology, CDH still carries a high mortality rate due to associated anomalies, particularly cardiac, chromosomal, and neurologic, with prematurity and low birth weight also affecting prognosis. Several recent reviews have documented an improved overall outcome for infants with CDH compared to 20 years ago with survival rates as high as 90%.

*Fetal interventions.* A clinical trial evaluating the efficacy of open surgical repair for fetuses with CDH demonstrated that the outcome of these infants was no better than infants cared for by conventional means. Thus, the optimism for open fetal surgical repair for CDH diminished, and other modalities were investigated.

An ongoing, prospective study from Europe (FETO group) evaluating fetal tracheal occlusion by inflatable balloon has presented preliminary results that are encouraging, but its limitations are the lack of a randomized design as well as standardized postnatal care.

**Test tasks**

**1. A newborn with a respiratory failure syndrome in a serious condition is hospitalized in a children's surgical department. The diagnosis was made during the examination: congenital emphysema of the upper lobe of the left lung. The method of treatment in this developmental disorder are:**

A. Observation tactics

It threatens the progressive deterioration of the child's condition

B. Conservative treatment

Does not provide the ability to eliminate tension in the pleural cavity

**C. Radical surgery**

The basis of the pathological condition of the child is the bloating of the parenchyma of the lungs, the tension in the pleural cavity, the displacement of the mediastinum, which can only be eliminated surgically. Radical surgery is to remove affected segments or lung particles

D. Puncture of the pleural cavity

In congenital emphysema, tension in the chest cavity is a consequence of hyperinflation of the parenchyma of the lung, so puncture does not lead to decompression

E. Drainage of the pleural cavity

In congenital emphysema, the tension in the chest cavity is a consequence of the burning of the parenchyma of the lung, so drainage will not lead to decompression

**2. In a child of 6 months of age there were symptoms of respiratory failure. The examination revealed a congenital cyst of the left lung, which was complicated by stress and located subplewrally within a single segment. Which operation will be most appropriate in this case?**

A. Lobectomy

The cyst is localized within the same segment, it is expedient to have an organ-saving operation

B. Segmental resection

The cyst is subpleural, it is possible to define its boundaries and to eliminate and thus preserve the parenchyma of the segment

C. Pneumonectomy

This operation leads to the loss of the entire lung and is inappropriate in a solitary bone

**D. Cystectomy**

The cyst is subpleved, it is possible to define its boundaries and to remove it. This is the most organically-safe operation.

E. Drainage of cyst

Not a radical surgical treatment. Can be used as a preparation for radical intervention for decompression of the cyst

**3. In a 3-month-old child with respiratory failure syndrome, the X-ray examination revealed increased transparency of the left pulmonary field, mediastinal hernia to the left, darkening of the right hemithorax, and the mediastinal displacement on the right with a tracheal infiltration in the upper chest aperture area. Pulmonary tissue in the right hemithorax is not determined. Identify the developmental defect that most likely corresponds to the described picture.**

A. Congenital emphysema of the left lung

Has a similar radiological picture, is included in the differential diagnostic series. Congenital emphysema is a hyperinflation of a lung area in the presence of another.

**B. Aplasia of the right lung**

The absence of signs of a collagen-like pulmonary tissue in the right hemithorax and a reshaped lung on the left indicates that the patient has only one lung - the left one.

C. Aplasia of the left lung

The redistribution is the parenchyma of the left lung, obviously it exists. The mediast displaced to the right - in the direction of the missing lung

D. Atelectasis of the right lung

Has a similar radiological picture, should conduct diagnostic diagnosis. It is possible to detect the airborne peptic parenchyma of the right lung.

E. Sequestration of the right lung

Usually, it does not lead to shift of the mediastinum. characterized by the presence of tumor-like formation in the cardio-diaphragmatic zone, more often on the left.

**4. Variant of the vascular ring, in which the highest mortality rate is observed:**

A. Pulmonary artery sling

Mortality without surgical treatment is about 90% during the first year of life, with surgical treatment - 5-45%.

B. Double aortic arch

The most lethal anomaly among the variants of the vascular ring is pulmonary artery sling

C. Aberrant right subclavian artery

The most lethal anomaly among the variants of the vascular ring is pulmonary artery sling

D. Right-sided aortic arch with left-sided arterial ligament

The most lethal anomaly among the variants of the vascular ring is pulmonary artery sling

E. Anomalies of the prickly trunk

The most lethal anomaly among the variants of the vascular ring is pulmonary artery sling

**5. A pathognomonic sign of the double aortic arch on the lateral esophagogram:**

A. Narrowing of the esophagus

This is a general term. In the case of a double aortic arch, the narrowing of esophagus has a specific configuration

**B. Defect of filling of the esophagus on the back wall with clear contours corresponding to the diameter of the aortic arc**

The esophagus and trachea are compressed in the vascular ring formed by the two arches of the aorta. The esophagogram shows the compression deformation of its lateral (in the anterior-posterior projection) and the rear (in the side projection) of the walls. The front wall of the esophagus is attached to the trachea.

C. Defect of filling of the esophagus on the front wall with clear contours corresponding to the diameter of the aortic arc

Double aortic arch leads to the compression of the esophagus on the lateral and posterior walls, which, respectively, are compression-deformed

D. Gastro-esophageal reflux

Has no specific signs of compression of the esophagus.

E. Suprastenotic enlargement of the esophagus

It is usually formed at the stenoses of the esophagus, which disturb its permeability and have a circular shape.

**6. A newborn child with respiratory failure syndrome has been diagnosed with congenital emphysema of the upper lobe of the left lung. The most frequent localization of congenital emphysema is:**

A. Right upper lobe

The left upper lobe is the most common localization of congenital emphysema

**B. Left upper lobe**

The left upper lobe is the most common localization of congenital emphysema

C. Right lower lobe

The left upper lobe is the most common localization of congenital emphysema

D. Left lower lobe

The left upper lobe is the most common localization of congenital emphysema

E. Right middle lobe

The left upper lobe is the most common localization of congenital emphysema

**7. A newborn child with respiratory failure syndrome has been diagnosed with congenital emphysema of the upper lobe of the left lung. Surgery is planned. Which operation is most appropriate?**

A. Upper right lobectomy

The lesions are localized in the upper lobe of the left lung

**B. Resection of apical segments of the left lung**

The most common is lesion of the apical segments of the left lung, therefore, it is expedient to have segmental resection of the lung

C. Upper left lobectomy

The most common is the damage to the apical segments of the left lung, so it is expedient to have segmental resection of the lung. Lobectomy is possible with damage to the entire particle

D. Left pneumonectomy

The lesions are localized within the particle. Pneumonectomy is contraindicated

E. Bronchoscopic occlusion of the bronchus of the affected particle

Will lead to atelectasis of the affected particle. Not a radical method. Possible for temporary removal of intracranial stress

**8. A 3-year-old child was under the dispensary supervision of a pulmonologist for frequent respiratory diseases. At regular examination the diagnosis was established: cystic hypoplasia of the lower part of the left lung. Method of choosing treatment for this pathology:**

**A. Surgical**

There is a threat of complications - the suppuration or tension of the affected lung. Surgical treatment is shown

B. Dispensary observation

There is a threat of complications - the suppuration or tension of the affected lung. Surgical treatment is shown

C. Surgical treatment is contraindicated

The lesions are localized within a single particle of lungs. No contraindications to surgery

D. Conservative treatment

It has a temporary anti-inflammatory effect. Can be used as a preparatory stage for the operation

E. Sanatorium-and-spa treatment

There is a risk of exacerbation of inflammation in the affected lung

**9. In a child aged 4 years, episodes of severe respiratory distress are periodically observed, the first of which is observed at 3 months of age after scheduled vaccination. At X-ray examination (Chest X-ray examination, cardioangiopulmonography), diagnosis was made: aplasia of the right lung. What therapeutic tactics should you choose?**

**A. Surgical - translocation of the dome of the diaphragm on the side of the defect**

Respiratory disorders are associated with the displacement of mediastinal organs, inflection and compression of the respiratory tract. The translocation of the diaphragm is a way to normalize the position and stabilize the mediastinum, eliminates the kinking and displacement of the trachea, contributes to its decompression.

B. Conservative treatment of respiratory diseases

Has only a temporary effect

C. Observation, prevention of respiratory diseases

Has no therapeutic effect

D. Surgical - removal of rudimentary lung

Does not match the pathogenesis of the defect

E. Sanatorium-and-spa treatment

Has only a temporary effect. Dangerous exacerbation of broncho-pulmonary inflammation and increased respiratory distress

**10. In what case at esophageal atresia is obligatory to perform gastrostomy?**

A. In all cases.

Performed only on certain indications

B. In cfse of anastomosis leakage

Executed, but not in all cases. Indications for gastrostomy depend on the degree of anastomosis leak

C. In fistulous form of esophageal atresia.

It is indicated only in the case of the impossibility of esophageal anastomosis

**D. In esophageal atresia without fistula**

Gastrostomy is a must-have primary surgery to ensure the child's ability to feed

E. With significant aspiration syndrome.

The genesis of aspiration syndrome should be determined. Its treatment is usually done without gastrostomy

**Situational tasks**

**Task 1**

A child 3 months of age has cough attacks during feeding. At the age of 1.5 months, there was a hospital treatment for pneumonia. Objectively: hypotrophy of II st.; Single moist rattles, shortness of breath, bloating. Exhaustion and diuresis are not affected.

1. What is the most probable diagnosis?

2. What diagnostic methods should be used to confirm the diagnosis in this case?

3. What is the therapeutic tactic in this case?

**Etalon response.**

1. Congenital isolated tracheoesophageal fistula.

2. Esophagography, tracheobronchoscopy with catheterization of fistula.

3. Surgical treatment - separation of the fistula from the cervical access.

**Task 2**

A child aged 9 months from the first days of life has a noisy breath, periodically noted distant wheezing, the involvement of auxiliary muscles in the act of breathing. Six times He had onsets of respiratory failure, four times the child was hospitalized to the intensive care unit. During the tracheoscopy at a distance of 3 cm from the vocal cords, the circular narrowing of the tracheal lumen as much as 60% of its lumen was detected. This bronchoscope tube could not be passed through this narrowing. Membranous part of the trachea in this area is absent. At examination of the optical system it was found that the narrowing of the lumen of the trachea reaches bifurcation, bronchial anatomy is not disturbed. The bronchial mucosa is moderately hyperemic.

1. What diagnosis is most probable?

2. What additional examination is necessary to determine the tactics of treatment?

3. What is the tactic of treatment in this case?

**Etalon response:**

1. Congenital long stenosis of the trachea (full of cartilaginous rings). Possible association with the lung artery loop (up to 50%)

2. Computed tomography of the chest with contrast enhancement

3. Therapeutic tactics are surgical: sliding tracheoplasty with artificial blood circulation (in association with the loop of the pulmonary artery - reimplantation of the left pulmonary artery).

**Task 3**

A child aged 4 months has an expiratory stridor, a rough barking cough with slight physical activity, attacks of breathlessness and cyanosis. The child was born with a body weight of 3200 g, the second day of life was operated on for esophageal atresia with distal tracheoesophageal fistula. The procedure consisted of the division of the fistula and end to end esophageal anastomosis. The child was extubated on the 12th day postoperatively. There was one unsuccessful attempt of tracheal extubation. The control esophagogram on 10th day after the operation showed a normal healing of the anastomosis of the esophagus.

1. What diagnosis is most probable?

2. What additional examination is necessary to determine the tactics of treatment?

3. What is the tactic of treatment in this case?

**Etalon response:**

1. Tracheomalacia is associated with esophageal atresia

2. Tracheobronchoscopy

3. Surgical treatment: aortoplexy, tracheoplasty or a combination of both operations.

**Task 4**

In a newborn child the condition is severe, shortness of breath, breathing on the left side of thorax is sharply weakened, percussion - box sound. On the plain chest X-ray the left hemithorax is of increased transparency with barely noticeable pulmonary pattern. Moderate displacement of the mediastinum to the right. In the lower section on the left there is a triangular shadow adjacent to the shadow of the heart.

1. What diagnosis can be established in this case?

2. What test methods should be used and for what purpose?

3. What is the surgical tactics?

**Etalon response:**

1. Congenital emphysema of the left lung

2. Computer tomography of the chest with intravenous contrast enhancement in order to clarify the localization of emphysema and differential diagnosis with aplasia or hypoplasia of the right lung.

3. Operation in an urgent order - resection of the affected segments of the left lung (more often S1-3)

**Task 5**

The child aged 1 year and 6 months since the age of about 1 month has stridor, frequent (up to 10 times a year) exacerbation of bronchitis, three times treated inpatient with a diagnosis of bronchopneumonia. During the examination, the child has a 10% weight loss, distant wheezing, and respiratory rate 35 minutes per minute. During auscultation markedly prolonged exhalation, scattered various-caliber moist rales are noted. On the review chest radiograph, the pulmonary pattern is enhanced on both sides, the right pulmonary field is more transparent than the left one. On the esophagogram in the lateral projection, there is a defect of filling the esophagus on the back wall in the projection of the aortic arch.

1. What diagnosis is most probable?

2. What is additional examination necessary to determine the therapeutic tactics?

3. Therapeutic tactics?

**Etalon response:**

1. Compression stenosis of the trachea by the vascular ring (a double arc of the aorta or right arc of the aorta)

2. Computed tomography with contrast or digital subtraction angiography

3. Surgical treatment: division of the vascular ring.

**Theoretical questions:**

1. What are the defects of trachea development you know?

**Etalon response:** The following are the defects of the trachea: trachea agenesis, tracheal atresia; congenital trachea stenoses, which are distributed into organic, compression and functional. Organic stenoses include complete cartilaginous rings. Such stenosis in turn is divided into the following types: generalized hypoplasia of the trachea, infundibular stenosis and segmental stenosis. Compression stenosis arises as a result of compression of the trachea by great vessels or tumors or cysts of the mediastinum. Functional stenoses include tracheomalacia.

1. What are the variants of compression stenosis of the trachea?

**Etalon response:**

- Compression stenosis as a result of the vascular ring: with a double artery of the aorta, right anterior artery aorta, aberrant right subclavian artery, loops of the pulmonary artery, anomalies of the branch and branching of the prickly trunk

- Compression stenosis due to compression of the trachea by mediastinal cysts (bronchogenic, enterogenic)

3. What operations are used in congenital tracheal stenosis?

**Etalon response:**

Depending on the length of the stenosis, slide tracheoplasty is used - with long stenosis (more than 1/3 of the length of the trachea), and segmental resection of the trachea - with short segmental stenosis (less than 1/3 of the length of the trachea)

1. What operations are used in compression stenosis of the trachea?

**Etalon response:** In the case of compression tracheal stenosis, the operation consists in decompression of the trachea, the method of operation depends on the variant of the defect that causes the stenosis. In the case of a vascular ring, the operation consists in division of the ring. In a case of double aortic arch one of the arches is divided, in the case of the right aortic arch the division of the ductal ligament is performed, in the case of the aberrant right subclavian artery the division of the right subclavian artery at the site of its origin from the aortic arch is performed, in the case of pulmonary artery sling reimplantation of the left pulmonary artery is the method of surgery, and with anomalies of the brachiocephalic trunk - anterior aortopexia is the operation of choice.

In the case of compression of the trachea by tumors or cysts of the mediastinum the surgery consists in removal of the tumor or cysts of the mediastinum.

1. What operations are used in tracheomalacia?

**Etalon response:** Depending on the mechanism of tracheal obstruction, aortopexy, tracheoplasty or a combination of them are used. Aortopexy is indicated in case of a deficiency of the cartilaginous framework and the prolapse of the anterior wall of the trachea, tracheoplasty - with a predominant prolapse of the back wall of the trachea. In the case of a combination of both mechanisms, aortopexy is combined with tracheoplasty.

1. What types of tracheomalacia are known to you?

**Etalon response:** Primary: idiopathic - a consequence of insufficient development of the cartilage of the trachea. More common in premature babies.

Primary associated with atresia of the esophagus and tracheo-esophageal bonyonus.

Secondary - is a consequence of compression of trachea by vessels or tumors, as well as the result of prolonged intubation or mechanical ventilation.

1. What are the possible variants of compressive stenosis of the trachea?

**Etalon response:** Esophageal atresia is distributed on the fistulous and non- fistulous forms. The following are attributed to fistulae forms: Atresia of the esophagus with distal tracheo-esophageal fistula, atresia of the esophagus with proximal tracheo-esophageal fistula, atresia of the esophagus with two fistulas, tracheo-esophageal fistula without atresia (H-fistula, congenital isolated tracheo-esophagus fistula).

8. Define the concept of congenital emphysema of the lungs

**Etalon response:** Congenital emphysema of the lungs is a rare developmental disorder characterized by hyperinflation and stretching of air normally formed parenchyma of segments or lobes, increases in their volume with a violation of lung ventilation and a decrease in the volume of blood flow

9. What pathogenetic factors lead to respiratory failure in the case of congenital emphysema of the lungs?

**Etalon response:**

- Hyperinflation of segments with pathological ventilation

- Formation of a mediastinal hernia, which compresses other parts of the lung and the opposite to the lung

- Emphysematous lungs do not participate in gas exchange due to violation of ventilation and blood circulation, ventilation of other lung units is reduced due to compression

- Shift of the mediastinum with tracheal kinking, compression of caval and pulmonary veins

- Compression of the trachea by the great vessels (arc of the aorta,brachiocephalic trunk)

- Violation of blood flow to the heart and outflow from the lungs

- Restrictions on chest and diaphragm excursions

10. What are the main anatomical differences of aplasia and pulmonary agenesis?

**Etalon response:**

Agenesis of the lungs. The trachea has no bifurcation and continues straight into the main lung bronchial tubes. The lung is absent, the existing lung forms a pulmonary hernia. Shifting the heart. The trachea is located near the middle line

Aplasia is light. The lung is absent or represented by a rudiment. Large (up to 2 cm) cuticle of the head bronchus. Bifurcation of trachea and carina developed. Dislocation of mediastinum, heart and pulmonary hernia. Transection of the trachea in the upper chest aperture. Bending and narrowing of the trachea in the bifurcation zone

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| **Etalon response:**Plain chest and abdominal radiograph, antero-posterior view. The mediastinal displacement to the right, multiple cellular enlightenments in the right hemithorax, the left dome of the diaphragm is not clearly visualized. Diagnosis: Left-sided diaphragmatic hernia |

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| А | Б |
| **Etalon response:** esophagograhy in anteroposterior and lateral views. The longitudinal esophagealaxis is interrupted, suprastenotic esophageal dilation. Indention of the lateral and posterior esophageal walls due to vascular compression. |

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| **Etalon response:** А – Chest and abdominal X-ray film of a neonate, anterio-posterior view. The tube is coiled in the blind-ending upper esophagus. Air outlining intestinal loops below the diaphragm confirms the existence of a distal tracheo-esophageal fistula. Diagnosis: Esophageal atresia with distal tracheo-esophageal fistula Б – Chest and abdominal X-ray film of a neonate, anterio-posterior view. The nasogastric tube is coiled in the upper esophageal pouch. Absence of air-filled abdominal intestinal loops suggests that there is no distal tracheo-esophageal fistula. Diagnosis: Esophageal atresia without distal tracheo-esophageal fistula (“Pure” esophageal atresia) |

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