PNEUMOLOGY - Part II

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AB – CLASSIFICATION - CONTROL

	Controlled asthma	**Partly controlled (Any measure)**	**Uncontrolled**
Daytimes symptoms	No (≤2 /week)	More than 2x/week	Three and more symptoms of partly controlled AB
Limitation of activity	None	Any	
Nocturnal symptoms	None	Any	
Need for relievers	No (≤2 /week)	More than 2x/week	
Lung function (PEF or FEV1)	Normal	< 80 % norm/personal best	

ASTHMA – TREATMENT – before 2019

Step 1	**Step 2**	**Step 3**	**Step 4**	**Step 5**
Inhalated B -agonists	RABA – on demand			
	One choice	***One choice***	***Add one or more drug***	***Add one or more drug***
	IKS low dose	IKS low dose + LABA	IKS middle or high dose + LABA	Oral corticosteroids
	Antileucotriens	IKS middle or high dose	Antileucotrienes	Anti - IgE
		IKS middle dose + antileucotrienes	Theophylline with prolonged release	
		IKS low dose+ theophylline with prolonged release		

Step long-term asthma therapy in children - GINA 2019

1. step	2. step	3. step	4. step	5. step
Děti od věku 12 let				
ICS+formoterol as reliever ev. ICS in case of SABA	Daily ICS or ICS in case of SABA or ICS + formoterol as reliever	Low-dose ICS + LABA or middle-dose ICS or ICS + LTRA	Middle-dose ICS+LABA or add tiotropium or add LTRA High-dose ICS	High-dose ICS+LABA or add tiotropium or add LTRA *Biologic therapy*
			Consultation of pneumologist´s	

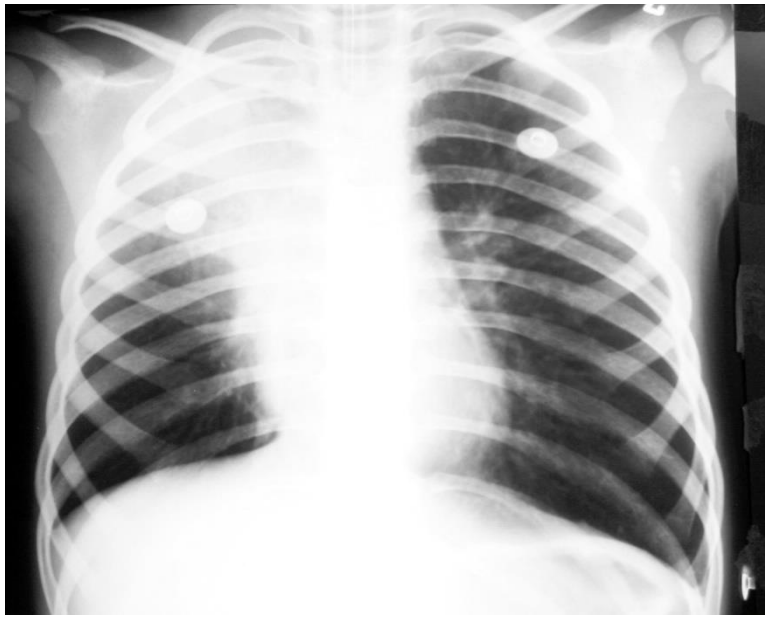
Centre for Severe Asthma:
Anti –IgE
Anti-IL5
Anti –IL 4/13
Bronchiál thermoplasty
Antimycotics
Macrolides

PNEUMONIA

- Infection of the lung parenchyma (respiratory bronchioli+alveoli+lung intersticium)

X pneumonitis – noninfectious inflammation

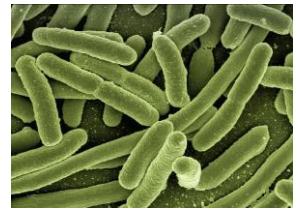
- Viral x bacterial x mycotic x parazitic
- Etiology and clinical picture different with age
- Baterial:
  1. Community acquired pneumonia
  2. Nosocomial pneumonia
  3. Pneumonia of immunosuppressed person



NEONATAL PNEUMONIA

- Clinical picture:
  1. Respiratory distress (tachypnoea, dyspnoea, retractions, grunting, cyanosis…)
  2. Nonspecific systemic signs – apnea, bradycardia, poor peripheral perfusion, temperature instability, hypotonia
- Etiology:
  - Bacterial infection – *Streptoc. agalactiae, E. coli, Listeria mono, Klebsiella pneu – risk of aspiration of amniotic fluid*
  - Viral – HSV, CMV, rubella, enteroviruses, RSV, parainfluenza, hMPV
  - Atypic agens – *Chlamydia trachomatis, Ureaplasma urealyticum*

ETIOLOGY ACCORDING TO AGE



Age		Infectious agens	
		Common	Rare
Newborns	0-5 days	Streptoc. agalactiae, E. coli, Listeria monocytogenes, Klebsiella pneumoniae	Haemophilus influenzae, Streptococcus pneumoniae, Ureaplasma urealyticum, Chlamydia trachomatis, anaerobes, CMV, HSV
	5-20 days	Dtto + nosocomial bacteria (Pseudomonas aeruginosa, Enterobacter, G-)	
Early infancy (3 weeks to 3 months)		Respirační viry (RSV, ADV, parainfl) Chlamydia trachomatis, Streptococcus pneumoniae	Ureaplasma urealyticum, Bordetella pertussis, Haemophilus infl., CMV, HSV, Str. pyogenes
From late infancy to preschool age (4m -5 Years)		Respiratory viruses, Strept. pneumoniae	Haemophilus influenzae Mycoplasma pneumoniae Chlamydophila pneumoniae, Moraxella catarrhalis, Mycob. TB
School age (above 5 years)		Strept. pneumoniae, Mycoplasma pneumoniae, Chlamydophila pneumoniae	Respiratory viruses, Haemophilus influenzae, Myc. TB, rarely Staph. aureus, Legionella pneumophilla

Community acquired pneumonia

= Pneumonia developed outside hospital or diagnosed within first 48 hours after admission to hospital of person – does not spend last 14 days in medical setting

Symptomatology:

1. *Common symptoms* - fever (usually above 38,5°C), cough, ev. tachypnoea, dyspnoea
2. *Atypical symptoms* – **abdominal pain** (CAVE! – may be dominant symptom), **vomiting**, **pain in shoulder and chest pain** = irritation of pleura - phrenic nerve, pleural pain increases with inspiration), backache, headache, meningeal syndrome within fever, grunting (rare symptom of severe clinical status)
3. *Oligo symptomatic course* – about 10 % of fever with unknown origin; in neonates apathy, apnoea, marks of sepsis

Physical examination

- Possible presence of dyspnoea (alar flaring of nose, retractions, work of accessory muscles) **and/or tachypnoea,** saturation of haemoglobin in peripheral blood
- Auscultation:
- **Crepitus or inspiratory fine crackles of small bubbles** and/or **dimished breathing** a/or **tubular breathing** – not highly specific but sensitive, common in other diagnosis – foreign body aspiration, pneumothorax, lung oedema, interstitial lung disease and further)
- Sounds develop in time – crepitus indux, crepitus redux

Pneumonia - diagnosis

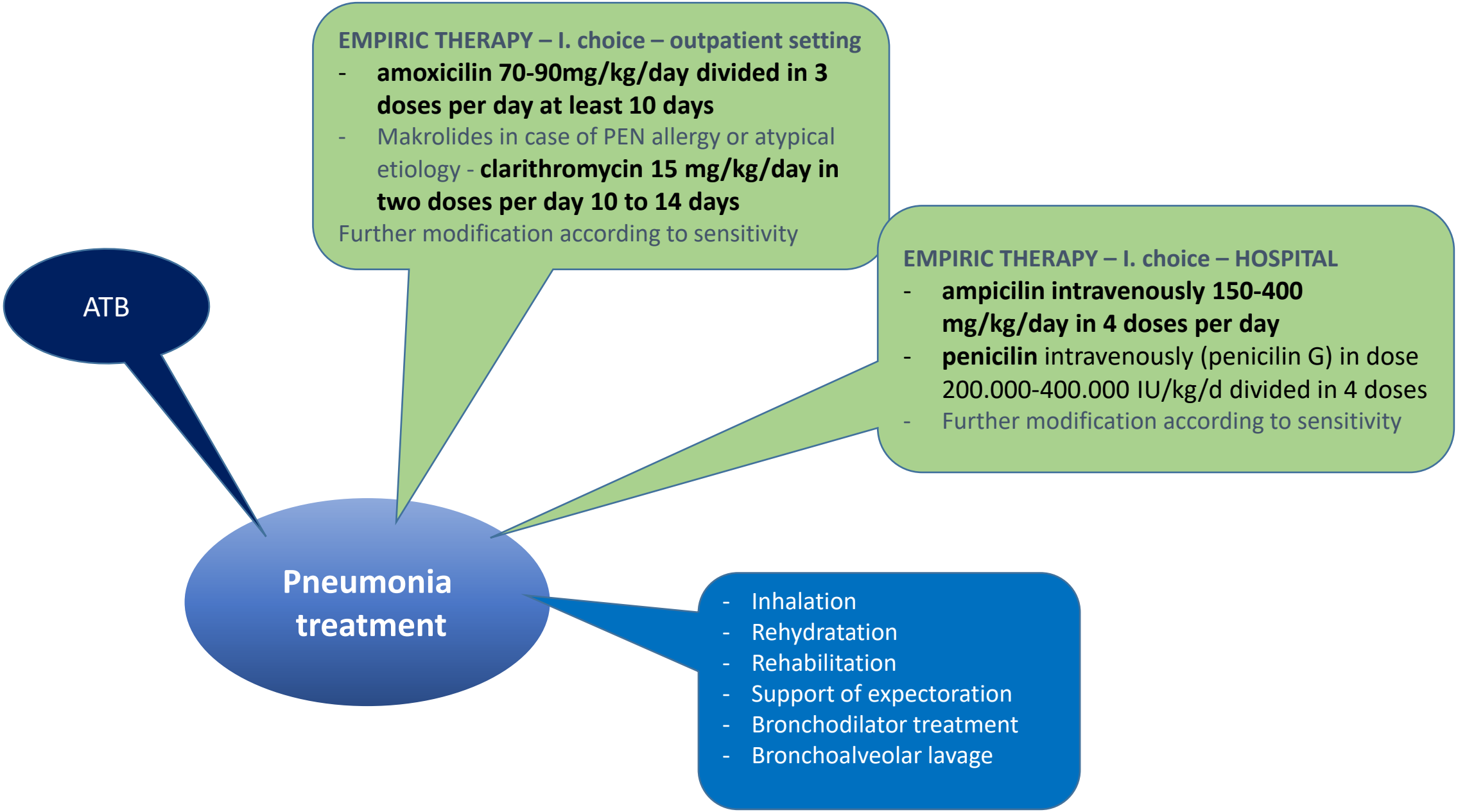
- Clinical symptoms + findings
- X-ray
  Etiology ?
- Laboratory tests
- Serology + microbiology

- Parameters of inflammation – CRP, sedimentation rate, procalcitonin

- Cultivation of mucosa without significance
- Sputum
- Hemoculture
- Urine - antigens (Legionella, Streptococcus pneu)
- Antigens in saliva –sérology (ADV, RSV, influenza, hMPV, Chlamydophila pn, Mycoplasma pn.)
- Serology (paired serum)

Bronchoscopy

- Suspected aspiration of foreign body
- Atelectasis
- Mucus plugging, samples for microbiological tests in case of severe pneumonia


ATB
EMPIRIC THERAPY – I. choice – outpatient setting
- amoxicilin 70-90mg/kg/day divided in 3 doses per day at least 10 days
- Makrolides in case of PEN allergy or atypical etiology - clarithromycin 15 mg/kg/day in two doses per day 10 to 14 days
Further modification according to sensitivity
EMPIRIC THERAPY – I. choice – HOSPITAL
- ampicilin intravenously 150-400 mg/kg/day in 4 doses per day
- penicilin intravenously (penicilin G) in dose 200.000-400.000 IU/kg/d divided in 4 doses
- Further modification according to sensitivity
Pneumonia treatment
- Inhalation
- Rehydratation
- Rehabilitation
- Support of expectoration
- Bronchodilator treatment
- Bronchoalveolar lavage

PNEUMONIA

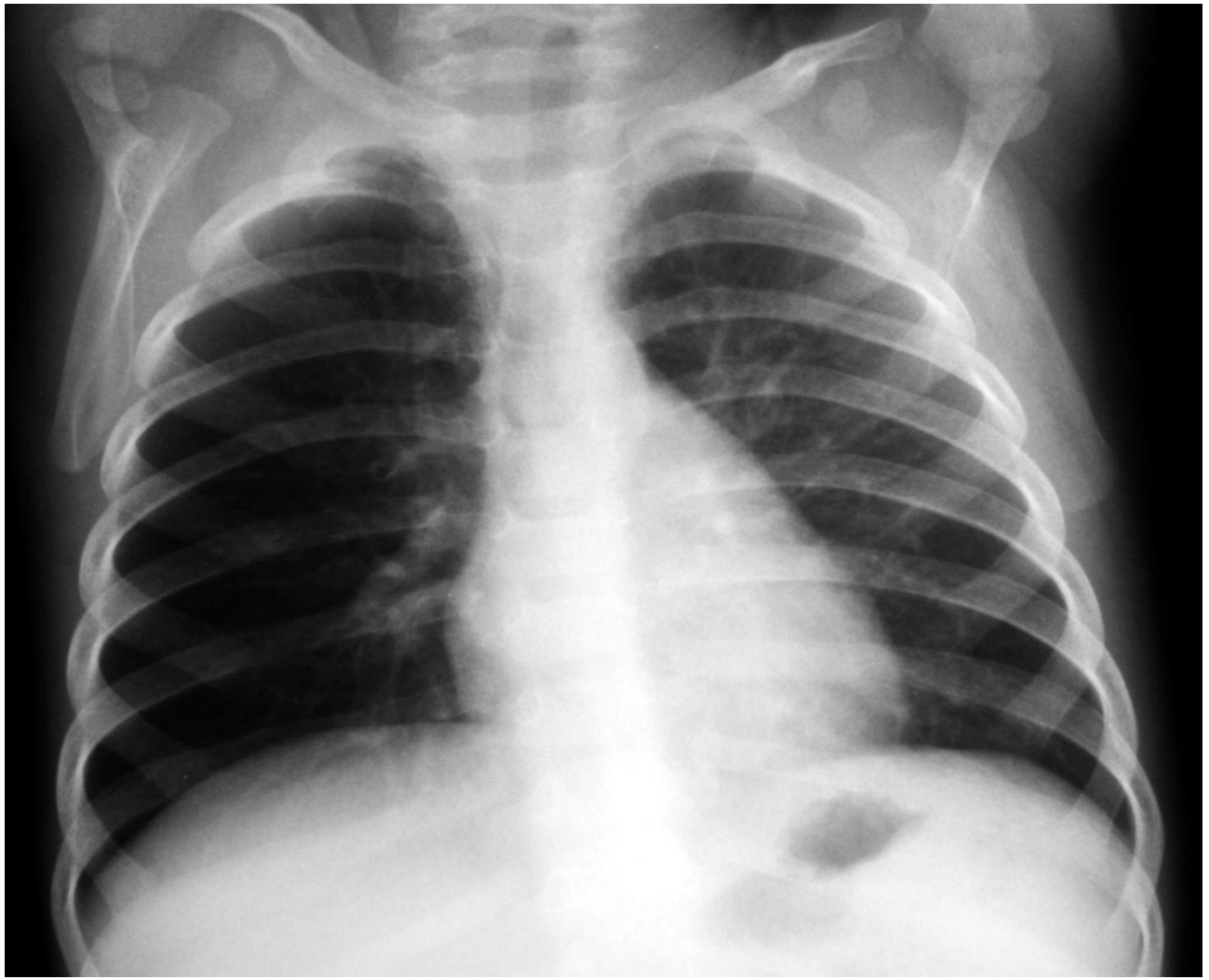
- CAP =community acquired x nosocomial
- RISK FACTORS :

1. Immunodeficiency
2. Chronic lung disease (asthma, cystic fibrosis, anomalies, bronchopulmonary dysplasia, alfa1-antitrypsine deficiency)
3. Immaturity
4. Severe course of pneumonia – hyposaturation, extrapulmonary symptoms – meningitis, arthritis…, severe X RAY- pleural effusion, infiltrates bilateral
5. Renal failure
6. Severe leucocytosis, leukopenia
7. Non – compliance of family

PNEUMONIA AND COMPLICATIONS

- Complicated pneumonia:
  - Atelectasis
  - Parapneumonic pleural effusion
  - Abscess
  - Necrotizing pneumonia
  - Incipient sepsis, metastatic infection
  - Pneumothorax
  - Bronchopleural fistula
- **Extrapulmonary complications**:
  - Search for signs of sepsis and metastatic bacterial infection
  - Haemolytic-uremic syndrome (HUS)
  - Syndrome of inadequate secretion of antidiuretic hormone (SIADH)

?



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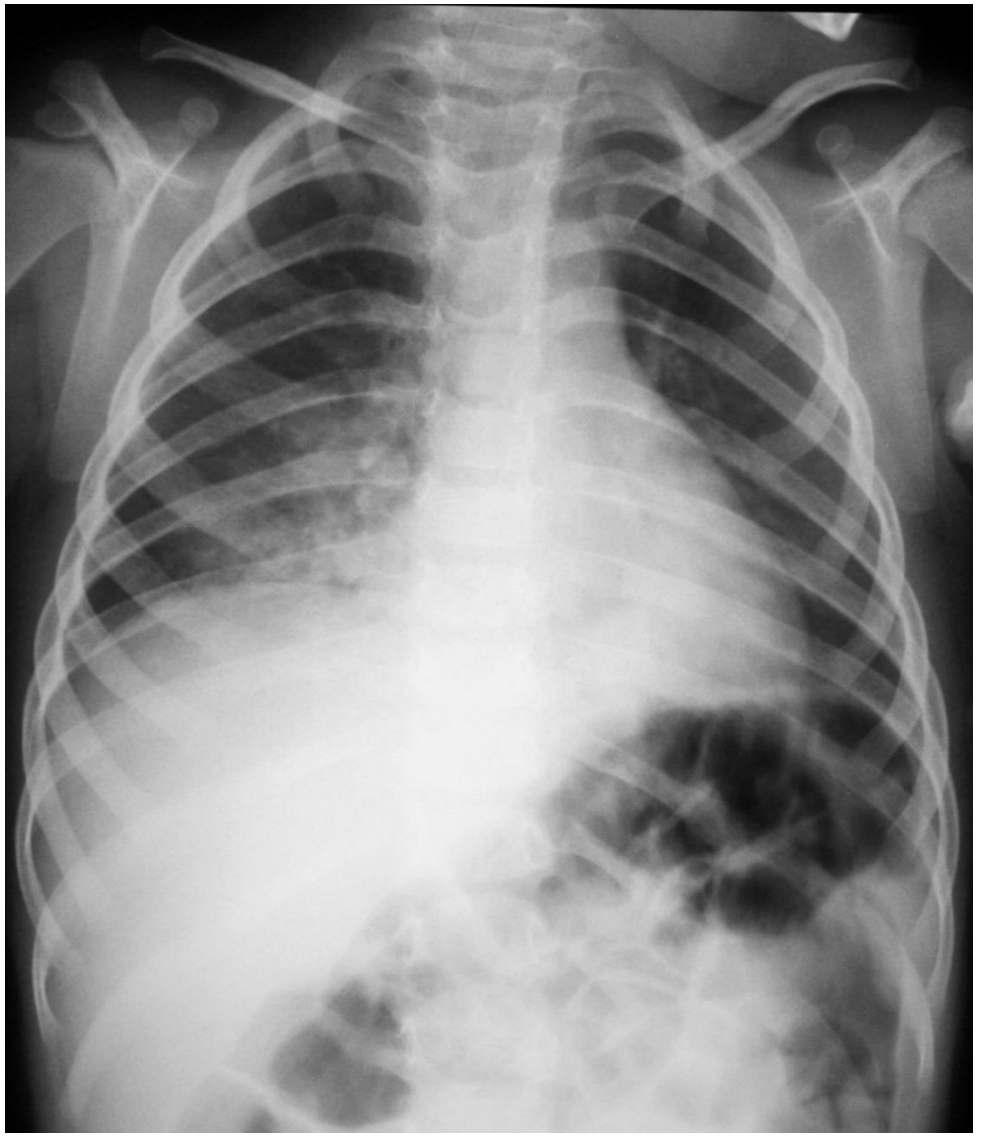


ASPIRATION OF FOREIGN BODY

- Subject ?
- Size?
- Amount?
- Age

RECURRENT PNEUMONIA

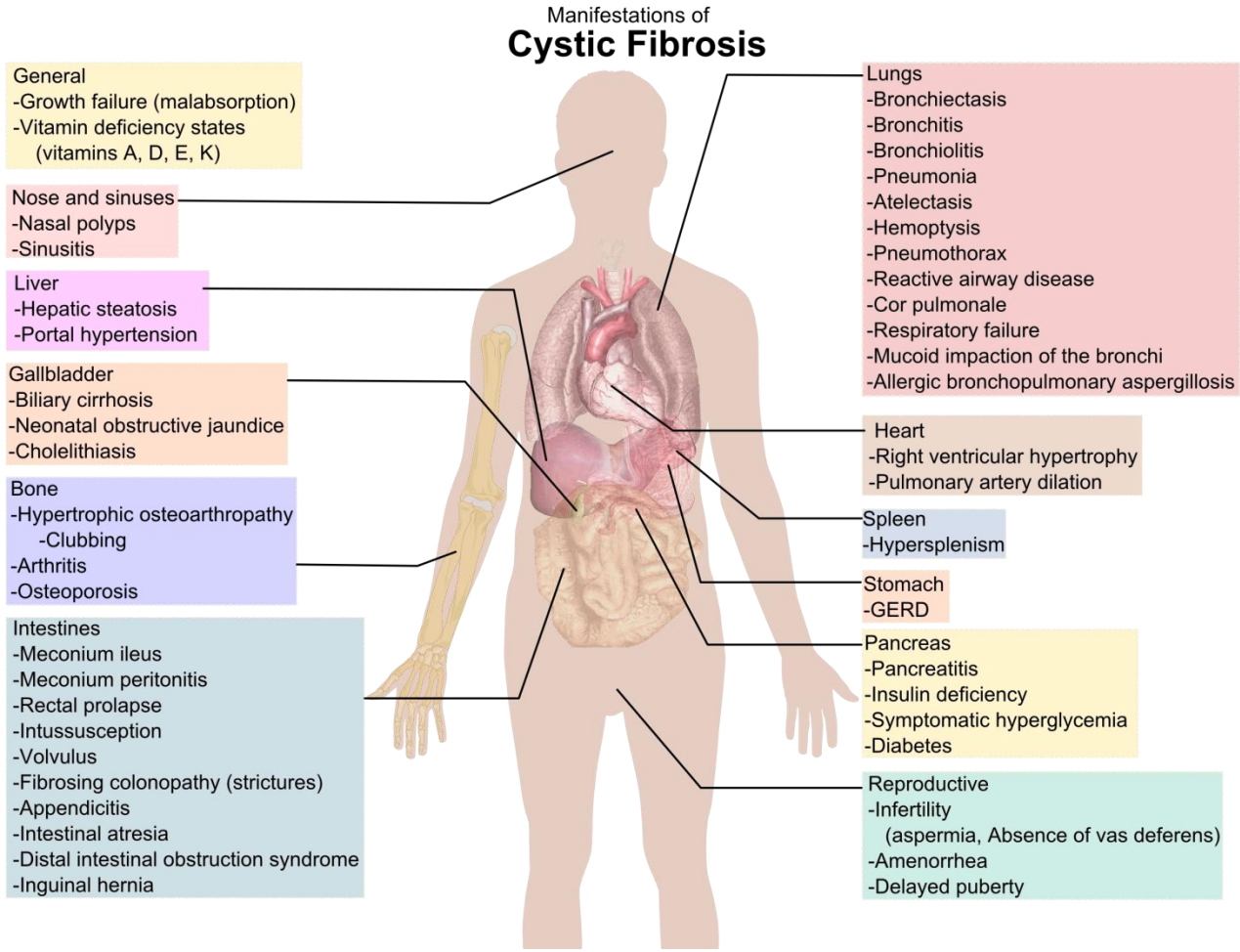
- Same localizations:
  1. Congenital anomaly of airways
  2. External or internal obstruction of airways
  3. Intralobar pulmonary sequestration

- Different localizations:
  1. Immunodeficiency
  2. Microaspirations, aspirations, GERD
  3. Primary ciliary dyskinezia

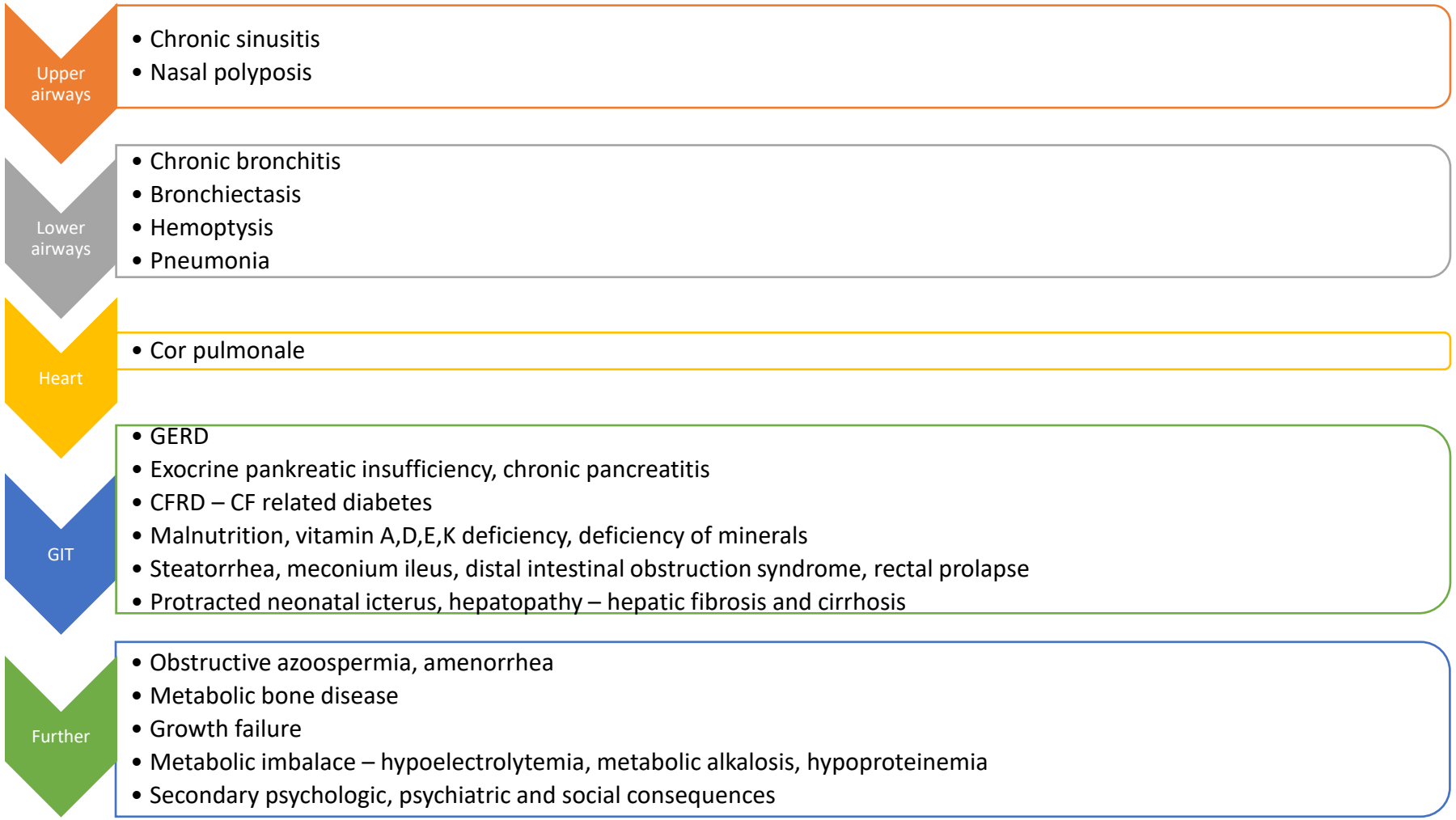
CYSTIC FIBROSIS

- Incidence: 1:2700-1:4000
- Autosomal recessive inheritance, neonatal screening
- CFTR (cystic fibrosis transmembrane conductance regulator protein)
  - 1600 known mutations, dominant F508del
- Multiorgan pathology
- Chronic sinopulmonary disease:
  - Chronic cough with mucous secretion, obstruction of airways
  - Bronchiectasis, atelectasis
  - Chronic pansinusitis, nasal polypes, recurrent pneumonia, wheezing, otitis
  - Colonization by microbi such as Pseudomonas, Burkholderia

CYSTIC FIBROSIS



MULTIORGAN PATHOLOGY



CYSTIC FIBROSIS

- Neonates:
  - Meconium ileus
  - Prolonged icterus
  - Failure to thrive – below birth weight - 1 mo of age
  - Hypoproteinemia with oedema
  - Metabolic crisis with hyponatremia, hypocalemia, hypochloremia a metabolic alkalosis
- Infants:
  - Failure to thrive in case of good or ravenous appetite
  - Steatorrhea – liquid stools are suspected mainly from cow milk allergy or celiac disease
  - Rectal prolapse

CYSTIC FIBROSIS

- OLDER CHILDREN:
  - Nutrition – below 3. perc. with weight
  - Recurrent sinusitis, nasal polypes
  - Chronic cough or recurrent bronchitis
  - Digital clubbing
- ADULTS
  - Obstructive azoospermia as an isolated symptom, sometimes connected with chronic sinusitis or mild respiratory symptoms

CYSTIC FIBROSIS: DIAGNOSIS

- Clinical symptoms, positive screening, family history
- Confirmation:

  Positive sweat test or 2 mutations of CFTR
- Neonatal screening - imunoreact. trypsinogen from dry drop – Guthrie test
  - positive =↑ test of frequent mutation CFTR
  - Pozit sweat test= diagnosis of CF x negat. – nosič

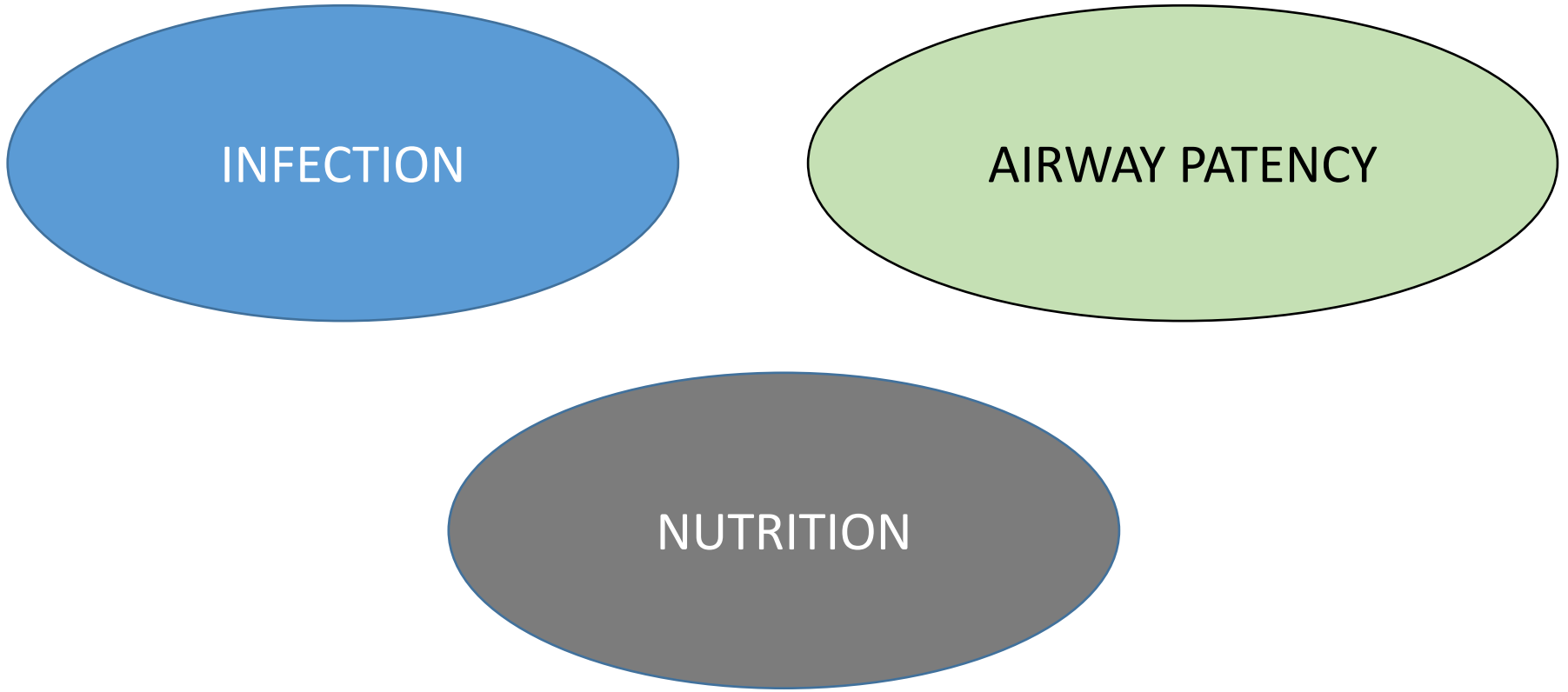
  CF-SPID – CF screen positive inconclusive diagnosis

- Sweat testing – collection by system Macroduct®
  - pilocarpine iontophoresis

  Cl >60 mmol/l 2x POSITIVE

  Cl - 30-59 mmol/l – borderline

CYSTIC FIBROSIS THERAPY



CYSTIC FIBROSIS THERAPY

- Infection:
  - Early treatment of infection – antibiotics
  - Surveillance of colonization and early eradication of microbi
  - Pseudomonas aerug., Burkholderia cepacia
- Airway patency:
  - Inhalation – hypertonic solution, mucolytics,
    human recombinant Dnase
    antibiotics
  - Respiratory physiotherapy
- Antiinflammatory treatment:
  - Azithromycin, ibuprofen in higher doses

CYSTIC FIBROSIS

- Daily energy requirement
  - 20-40 % higher than in healthy children
  - 35-45 % of energy intake by lipids, mainly vegetable fats
- MALNUTRITION:
  - Decreased resorption of nutrients (pancreatic insufficiency) – substitution of pancreatic enzymes
  - Higher energy consumption (respiratory muscles, expectoration)
  - Hepatopathy (not all patients)
  - Anorexia x hunger
  - Chronic infection

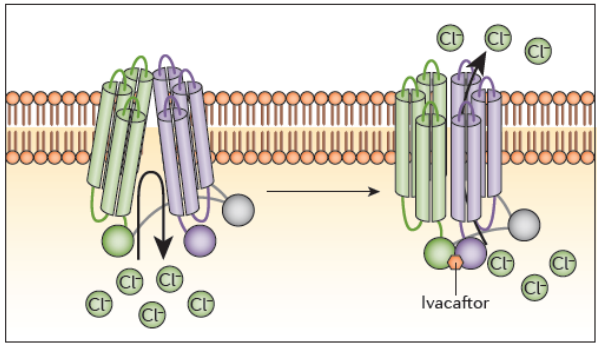
CF - PROGNOSIS

- 50 years ago – life expectancy ~ 2 years
- Present – about 50 % of patients survive 25 - 30 y of life
- Current neonates may survive more than 40 years

Presence and future of CF ?

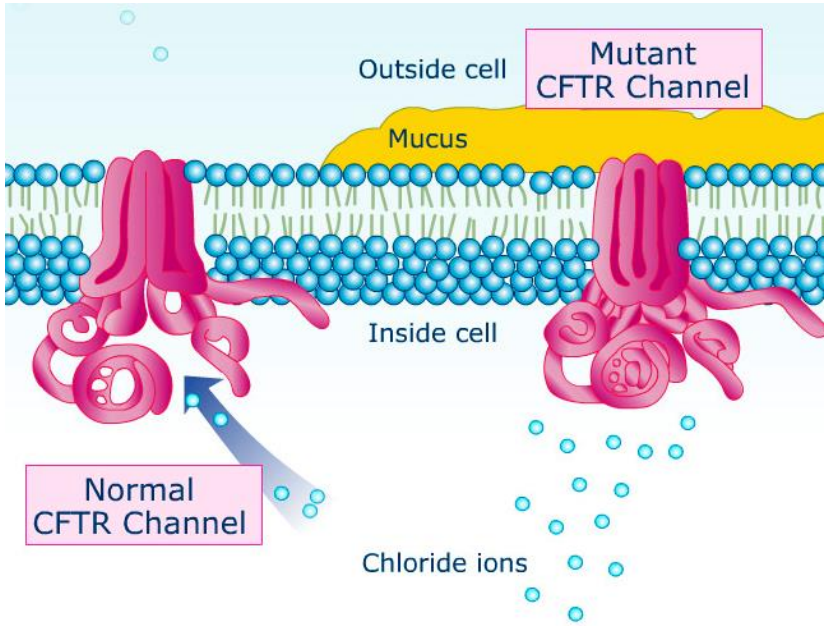
- Therapy aimed at specific molecular defect
- Functional classification of CFTR gene mutation

Třída	I	II	III	IV	V	VI
Typ of CFTR dysfunction	Failure of synthesis	Failure of intracellular transport and processing	Failure of activation	↓conductivity of channel	↓ synthesis	↓ stabilisation of protein
Example	G542X	F508 del N133K	G551D	R347P D1152H R117H	3849+10 kb C→T	



CF - pitfall

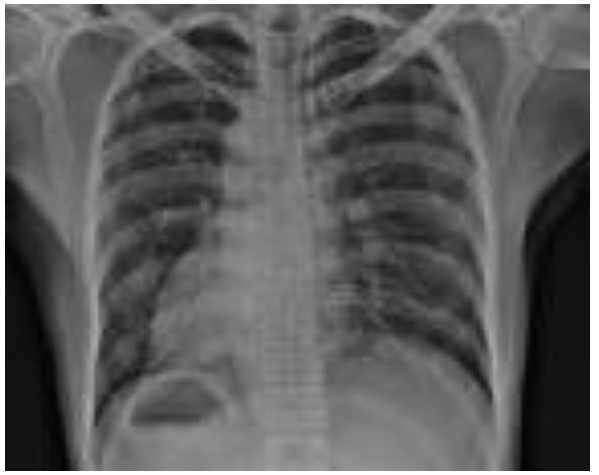
- CFTR - cystic fibrosis transmembrane conductance regulator protein
  - Regulation of chloride conductance
  - Inability to secrete salt and secondary to secret water + excessive reabsorption of salt and water
  - Viscous desiccated secretion



- Salty skin baby

http://prep-pg.blogspot.cz/2012/04/revolution-in-management-of-cystic.html

PRIMARY CILIARY DYSKINESIS



- AR
- 1:10.000-1:40.000
- Ciliar dysmotility of respiratory and sexual systém and malfunction of nodal cilia involved in embryogenesis:
  - Decreased mucociliar clearance
  - Decreased fertility (man – infertility, female ↑ risk of extrauterine gravidity)
  - Incorrect embryogenesis – situs viscerum inversus – in same patients, congential heart disease
- Kartagener´s syndrome - chron. sinusitis+bronchiectasis+chronic cough
- Clinical picture:
  - RDS in mature newborn, neonatal rhinitis, chronic rhinosinusitis, chronic secretoric otitis
- Diagnosis: nasal FeNO, examination of nasal cilia – high speed electron videomicroscopy

https://radiopaedia.org/articles/situs-inversus

TUBERCULOSIS

- Mycobacterium tuberculosis complex
  - Mycobacterium tuberculosis
  - Mycobacterium bovis
  - Mycobacterium bovis BCG
  - Mycobacterium africanum
  - Mycobacterium microti
  - Mycobacterium canetti

- Nontuberculous Mycobacteria
  - More than 100 kinds with different pathogenicity

TUBERCULOSIS

- Incidence – 3.56/100.000 (2022) – 7.25/100000 Prague
- 88 % pulmonary
  - 19 children < 19 years (2022)

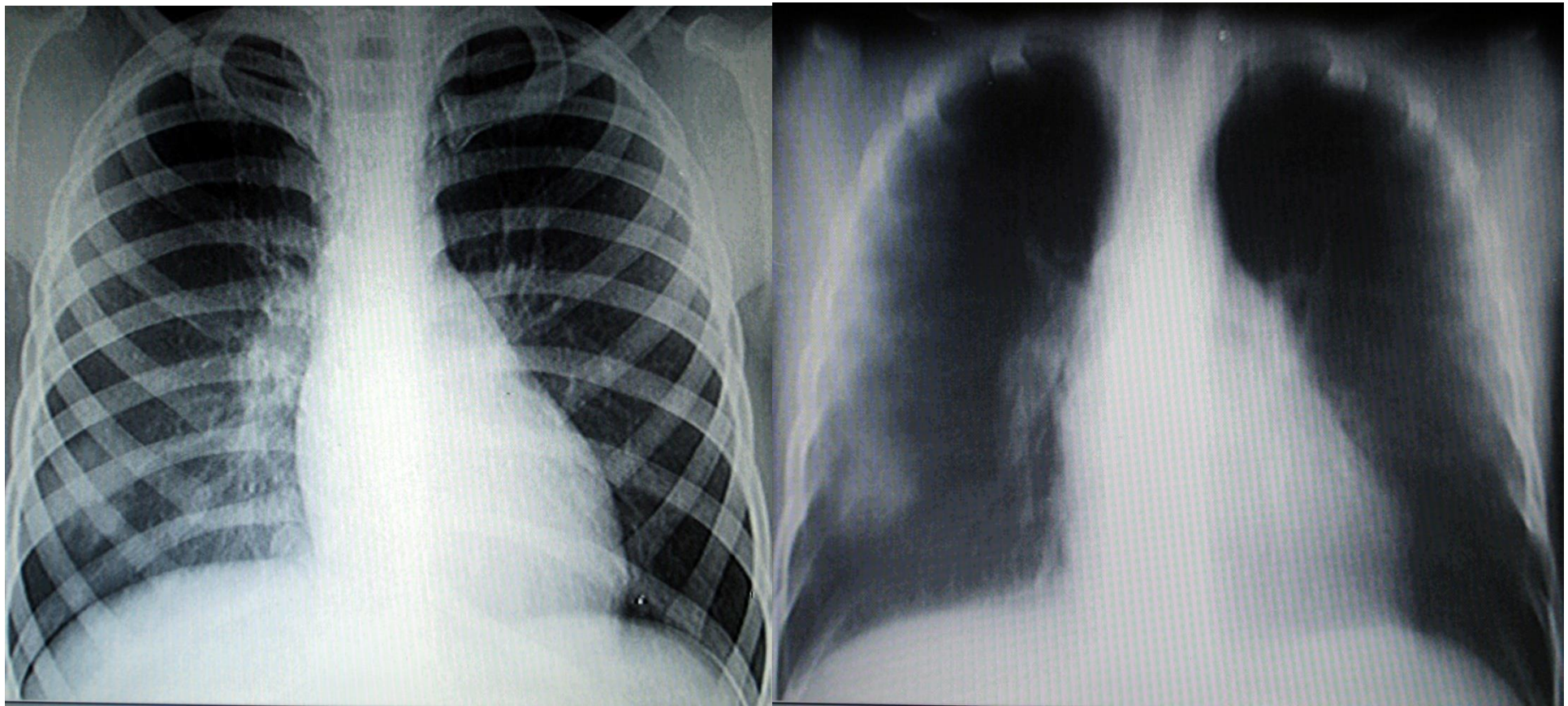
- Latent form x Active TBC

(noninfectious)

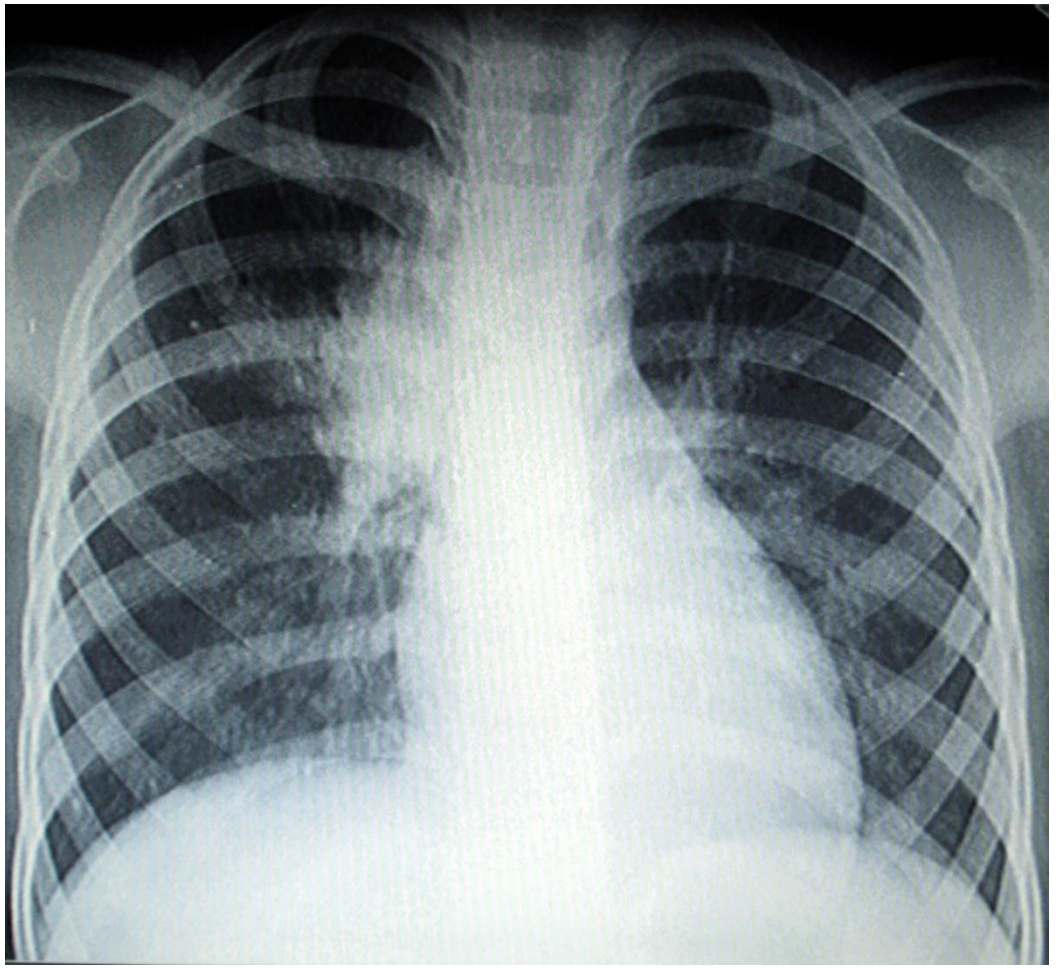
- Primary complex = primary lung lesion + lymph node
  - No clinical and laboratory symptoms
- Postprimary tuberculosis
  - Cough, night sweats, subfebrile, fatigue, anorexia, weight loss
- Miliary TBC

TUBERCULOSIS

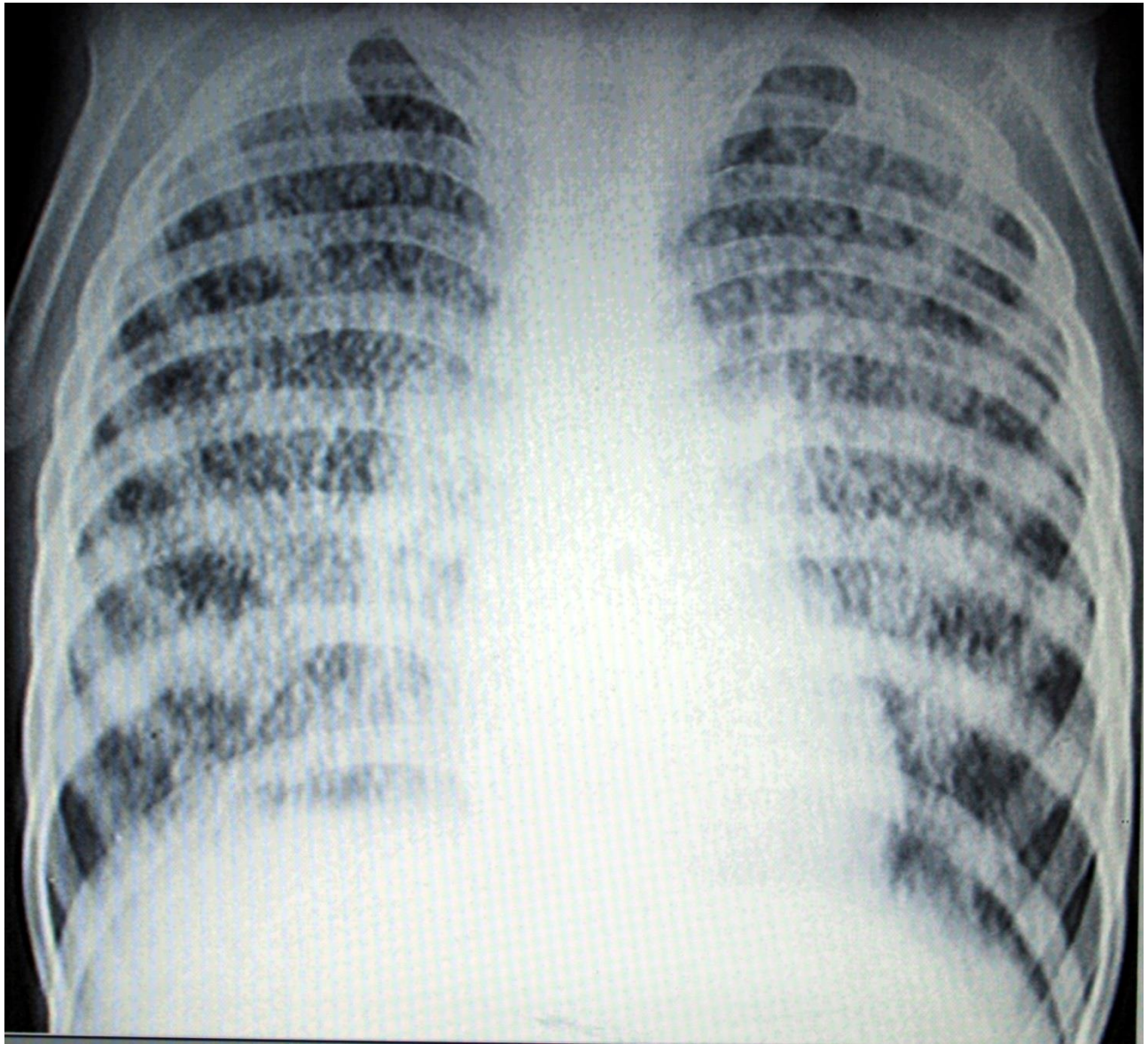
Thorax



TUBERCULOSIS



TUBERCULOSIS



ECDC, 2017

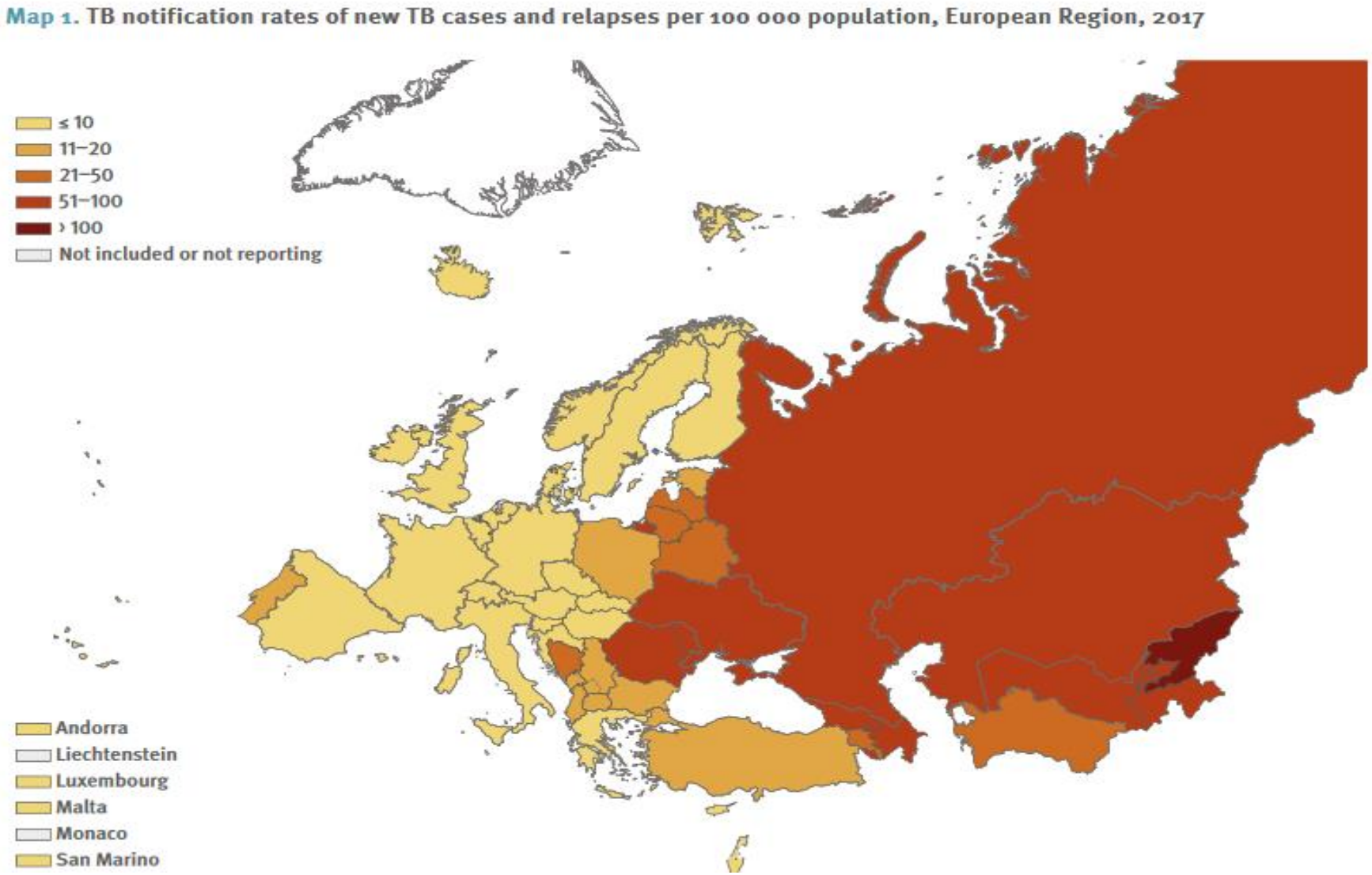
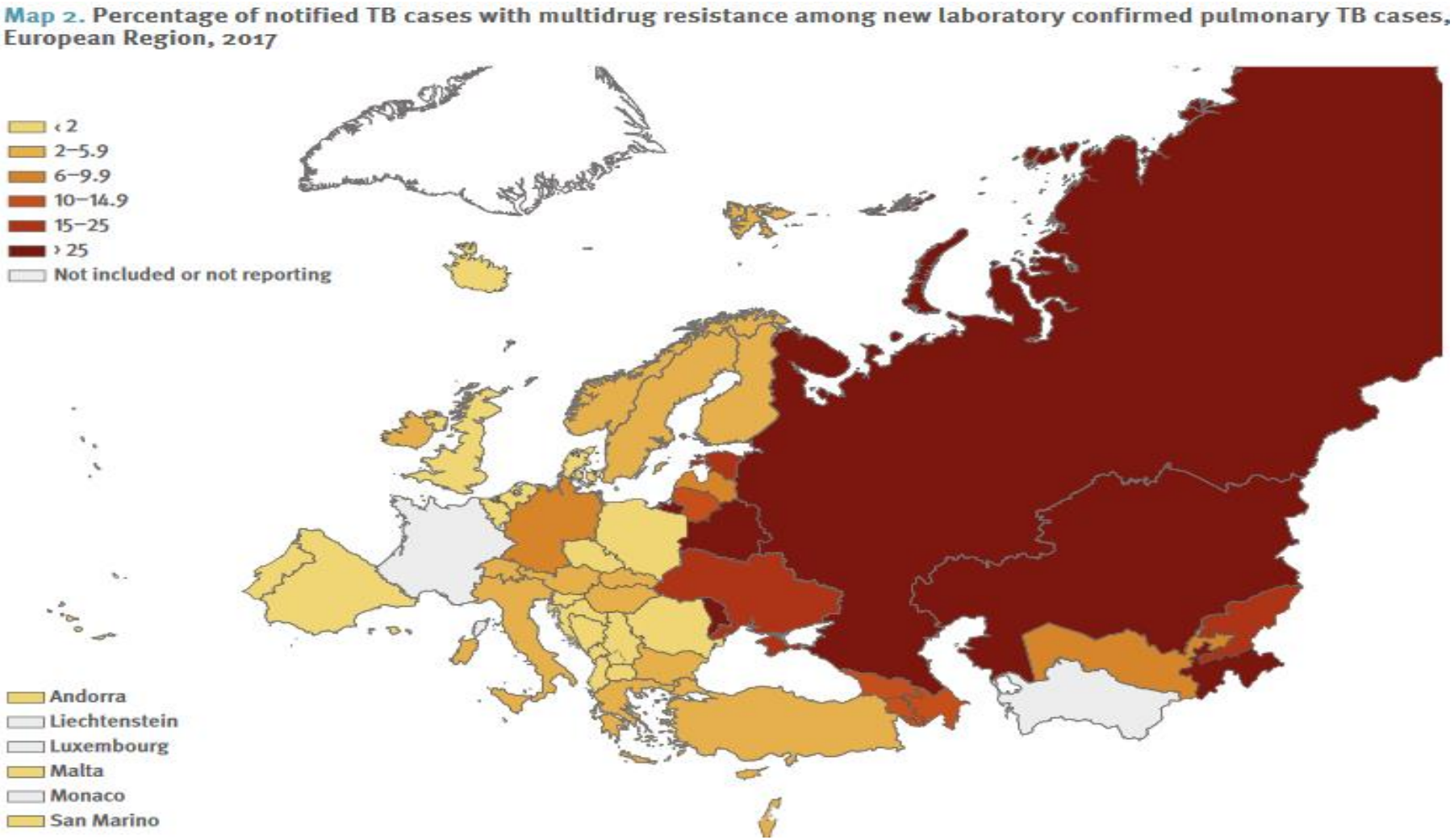
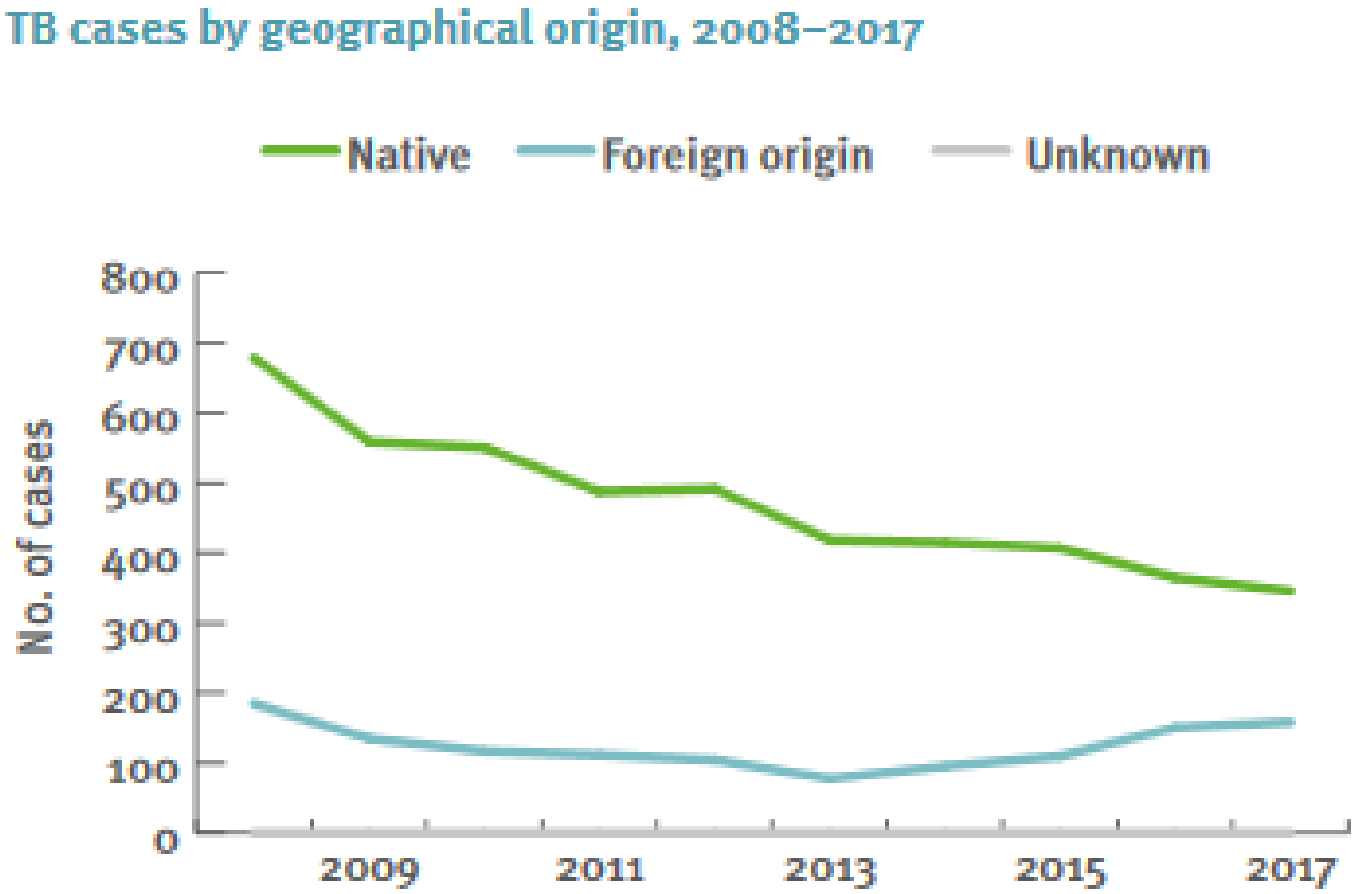

Map 1. TB notification rates of new TB cases and relapses per 100 000 population, European Region, 2017
≤ 10
11–20
21–50
51–100
› 100
Not included or not reporting
Andorra
Liechtenstein
Luxembourg
Malta
Monaco
San Marino

Fig. 5. TB notifications per 100 000 population by year of reporting, EU/EEA, 1995–2017

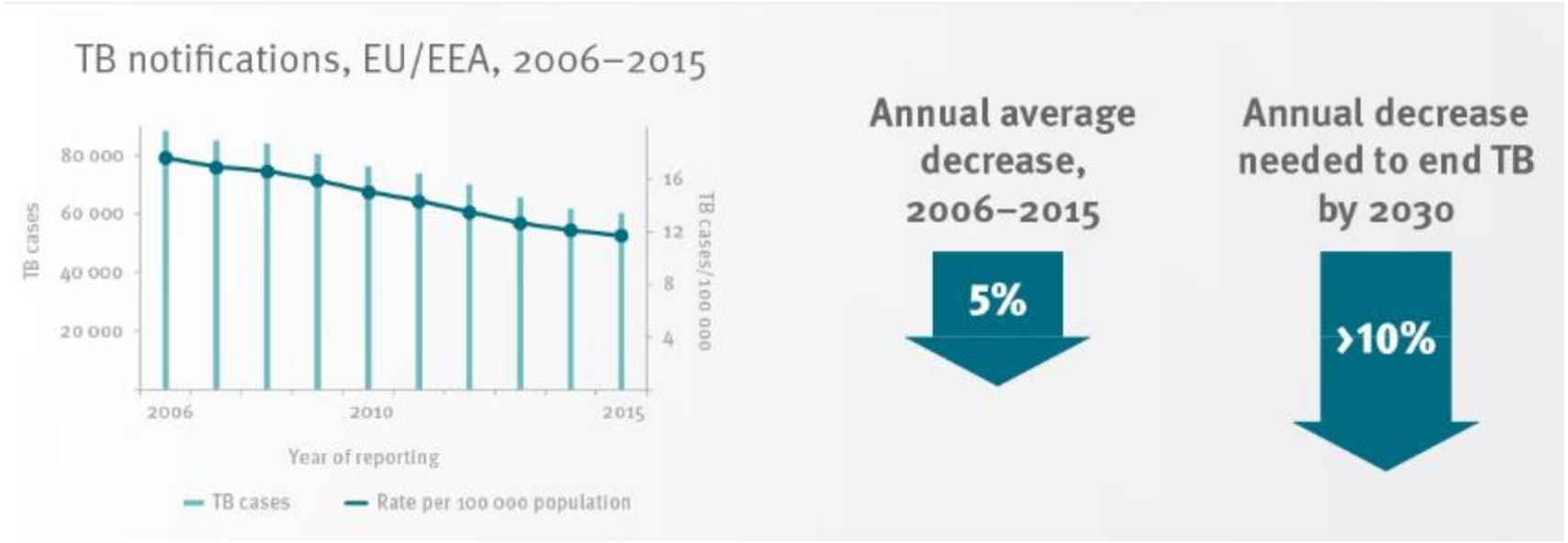
ECDC, 2017


Map 2. Percentage of notified TB cases with multidrug resistance among new laboratory confirmed pulmonary TB cases, European Region, 2017
< 2
2–5.9
6–9.9
10–14.9
15–25
> 25
Not included or not reporting
Andorra
Liechtenstein
Luxembourg
Malta
Monaco
San Marino

Proportion of foreigners, Czech



European Centre for Disease Prevention and Control. TB surveillance and monitoring in Europe, 2019

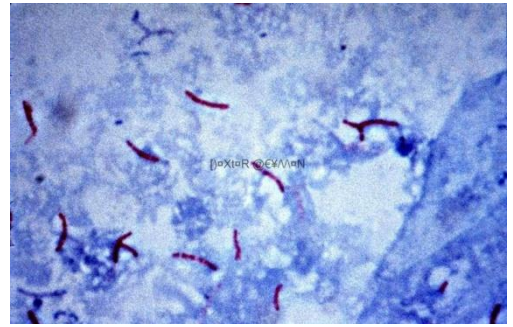


European Centre for Disease Prevention and Control. TB surveillance and monitoring in Europe, 2017

TBC - DIAGNOSIS

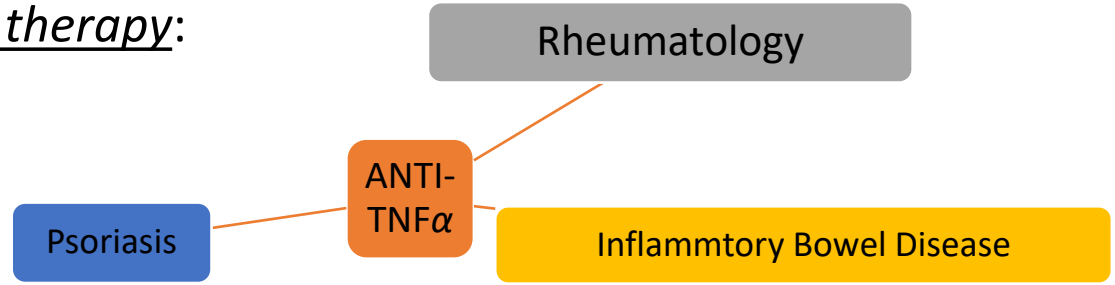
- Suspicion – clinical or epidemiology
- Possible contact
- X-RAY
- Tuberculin test – 2 TU /0.1 ml Manoux II
  - Negative - induration < 5 mm
  - Pozitive - > 6 mm: ≤ 10 mm postvaccination
    > 10 mm – postinfectious
    > 15 susp. active infection

- Quantiferron TB Gold – IFN gamma
- PCR – high false negativity, false positivity
- Microscopy – Ziehl-Neelsen, fluorescence – sputum, gastric lavage, laryngeal swab
- Cultivation – sensitive – duration 6 w
  - Faster method – 2 weeks



ANTI-TNFα THERAPY and TB

- *<u>Indication of anti-TNF therapy</u>*:



- ***TNF α*** = production of granuloma

  Increase ability o macrophages to kill mycobacterium

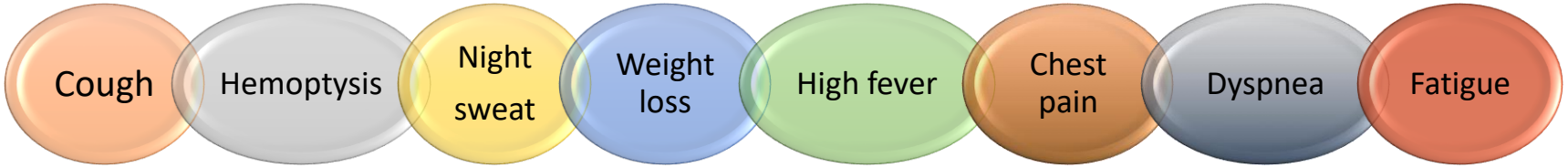
X inhibition of TNFα = increase risk of TB reactivation or new infection - about 4 – 8x[Vencovsky J. et al, Ces revmatol, 2009]

✓

DIAGNOSIS of LTBI (1)

!!! Always exclude active infection !!!:

- History (country of origin, travelling, contact with TB, history of TB infection, malnutrition, imunosupresive treatment, HIV status)
- Symptoms – aimed questions:



Most specific in developing countries according to EBM Marais et al, Pediatrics, 2006:

1. *Cough above 2 weeks*
2. *Failure to thrive in last 3 months*
3. *Fatigue*

- Chest X-ray

DIAGNOSIS of LTBI (2)

1) <u>In vivo</u> = tuberculine skin test (TST = tuberculin sensitivity test) = Mantoux II test 2 IU of tuberculine in 0,1 ml – dorsal left forearm intradermally – result in 48 – 72 hours

2) <u>In vitro</u> = IGRA tests (Interferron Gamma Release Assays)

= blood tests evalueting presence of specific reactive T lymphocytes that produce *interferon- γ* after specific interaction with antigen of mycobacterium

- **QuantiFERON®-TB Gold, Gold In-Tube (from 2006)**
- **T-SPOT.TB™**

✓ Not for differentiation of active and latent infection

<u>Always necessity to exclude active TB infection!!!</u>

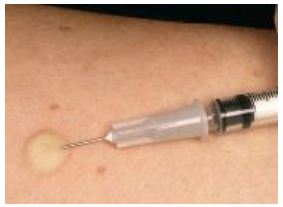
✓ Not for prediction of development of active infection

✓ Not for differentiation of reinfection and reactivation

TUBERCULIN SENSITIVITY TEST (TST)

Infiltration=induration Canadian Respiratory Guidelines 2013:

- < 5 mm negative reaction – sensitivity 98 %, specificity 60%
- >10 mm - sensitivity 90%, specificity >95%
- >15 mm sensitivity 60%, specificity >95%



Interpretation according to ATS:

- Positive > 5 mm:
  - ✓ Imunosupresion (above 15 mg of prednisone more than month)
  - ✓ Biologics
  - ✓ Transplant recipient
- Positive > 10 mm:
  - children < 5 let
  - Children and adolescents in contact with TB

Above 5 mm =positive, in vaccineted person 6-15 mm coulg be postvaccinated reaction

Infiltratrion=induration[Křepela et al]:

- < 5 mm – negative reaction
- 6-10 mm – usually postaccinated reaction
- >15 mm – positive
  - ➢ ***In children below 5 years >10 mm***

Infiltration=induration:

- < 5 mm – negative reaction

SKIN TUBERCULIN TEST (TST)

Advantages

- Cheap
- Highly sppecific in nonvaccinated population (up to 95%)
- Preferred when repeated testing is planned - screening
- Prefferd in developing countries (according to WHO)

Disadvantages

- Two visits (result 48-72 h)
- Subjective evaluation, risk of invalid application
- Different interpretation
- ↑ increased false positivity:

1. BCG vaccination
2. Infection by nontuberculous mycobacteria (NTM)

- ↑ falce negativity:

1. Imunosupresion therapy (Prednison >= 15mg/day>2-4 weeks)
2. imunodeficiency (HIV), systemic disease, malnutrition, active TB, elderly, infants below 6 months, viral infection, vaccination

- Booster efect after repeated application

IGRA TESTS

Advantages

- Highly specific in vaccinated population (above 95% for LTBI, (independent on BCG vaccination)
- Independent on infection by NTM (except for Myc. kansasii and marinum)
- ↑ sensitivity in case of imunosupresion, imunodeficiency or systemic inflammatory disease
- Decrease numbers of patients indicated to chemoprophylaxis

Disadvantages

- Expensive
- More exacting on laboratory and personal facilities
- Bigger blood sample (children)
- Time limit for processing
- Risk of false negativity in active infection, malnutrition, severe acute infection

TUBERCULOSIS - DIAGNOSIS

1. TBC verified by bacteriol. or histol. methods:
- Microskopic license of mycobacteria or positive cultivation of biological material
- Histological proof of tuberculous nodules or exsudates

2. TBC not verified by bacter. or histol.:
- Dg. Suspected from anamnesis, RTG, Mx II, cytology, PCR, BACTEC
- Dg. without verification of mycobacterium

3. Miliary TBC

4. TBC of CNS

5. TBC of other organs

TUBERCULOSIS - THERAPY

- 2 phases: Initial – 2 mo
  Follow – 6- 10 mo
- Combination of drugs – prevent resistance
- Once daily – REGULAR APPLICATION
- Risk of toxicity

ISONIASID (+pyridoxin), RIFAMPICIN, ETHAMBUTOL,PYRAZINAMID

STREPTOMYCIN

X

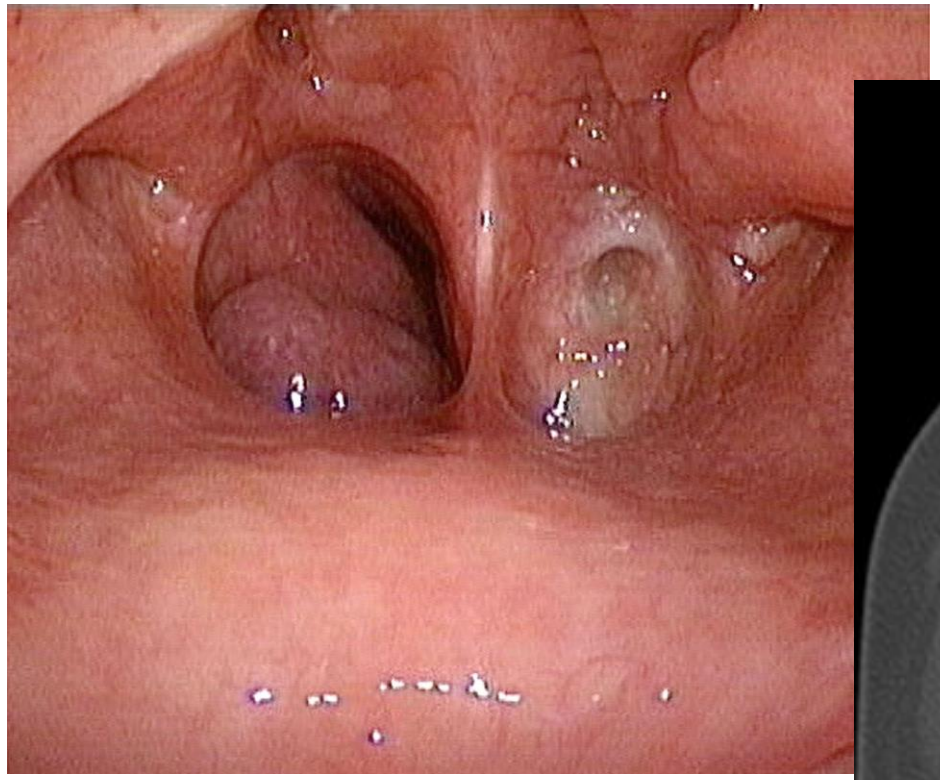
- Chemoprofylaxis - isoniasid 5mg/kg/d – 6 mo – monother.
  - Positive Mantoux in nonvaccinated child with negative X RAY
  - Contact with active TBC and negative X-RAY
  - Conversion of Mantoux II from postavaccin. to postinfectious reaction
  - Negative X-RAY and Mantoux II > 15 mm (child below 5 y >10 mm)

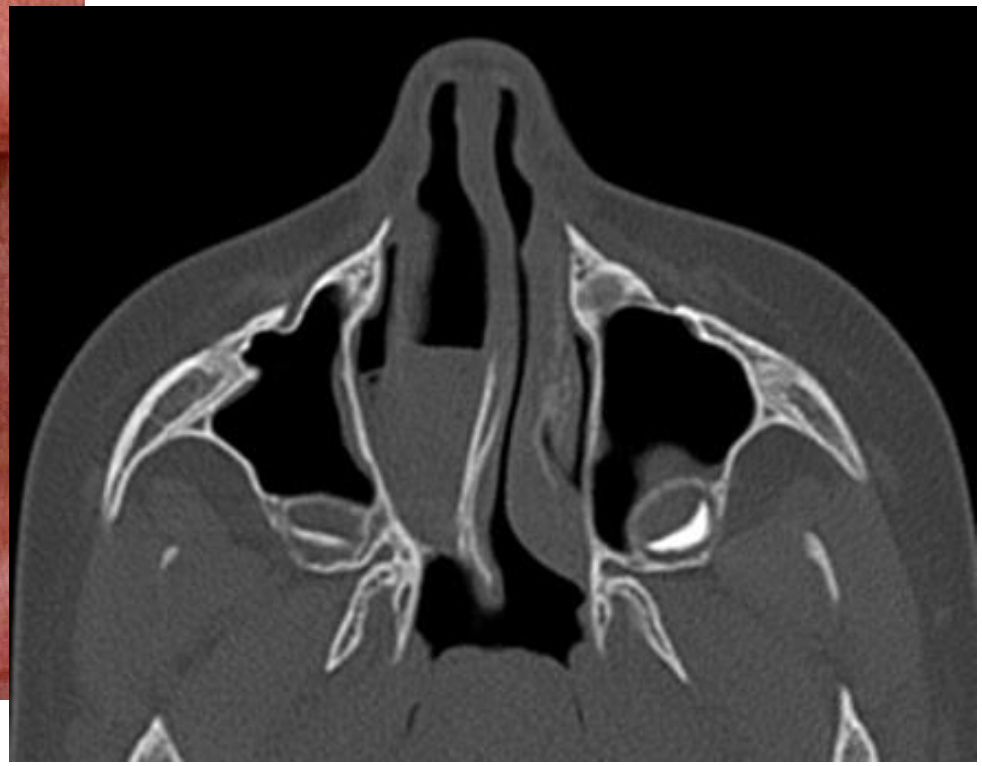
TUBERCULOSIS

- LIVE vaccine!!!!! – Mycobacterium bovis
- CZ – 2010 : Vaccination of risk group – age < 6 w

  - voluntary – after basic vaccines – negative tuberculin test
- Risk of generalization of BCG infection in SCID
- VACCINATION – protection against:
  - Systemic forms of TBC
  - Atypic TBC – avium…

Congenital disorders





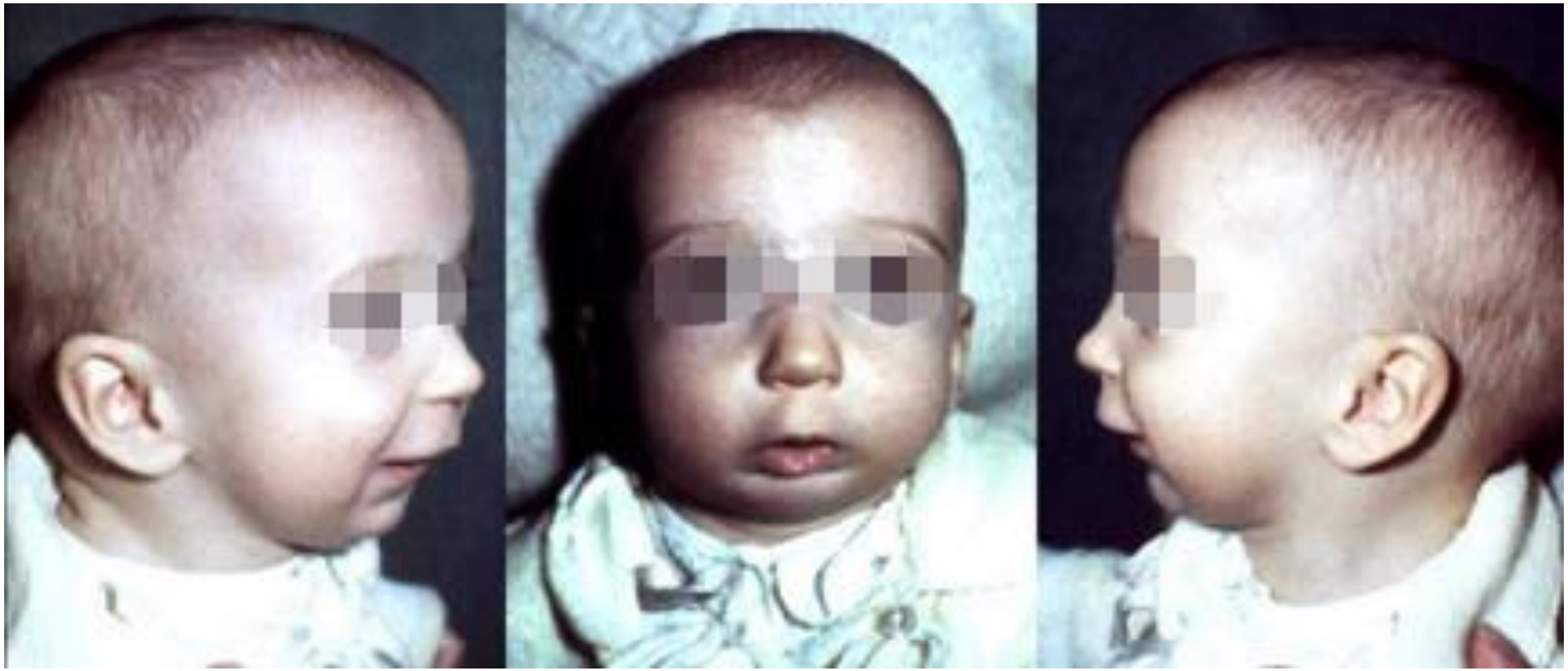
CONGENITAL DISORDERS OF THE NOSE

- Nasal hypoplasia
- Arhinia
- Supernumerary teeth
- Congenital nasolacrimal duct obstruction
- Choanal atresia
- Congenital defects of the nasal septum
- Pyriform aperture stenosis
- Congenital midline nasal masses – dermoids, gliomas, encephaloceles

CHOANAL ATRESIA

- 1:7000
- Unilateral x bilateral
- Bony 90 % x membranous
- 50-70% association with other anomalies
- 10-20% → the CHARGE syndrome

  Coloboma **H**eart disease **A**tresia choanae **R**etarded growth **G**enital anomalies **E**ar anomalies
- Clin. manif. – variable, cyanosis relieved by crying pláčem x during sucking
- Dg – catheter, fiberoptic rhinoscopy, HRCT
- Therapy – intubation, oral airway



Congenital facial anomalies

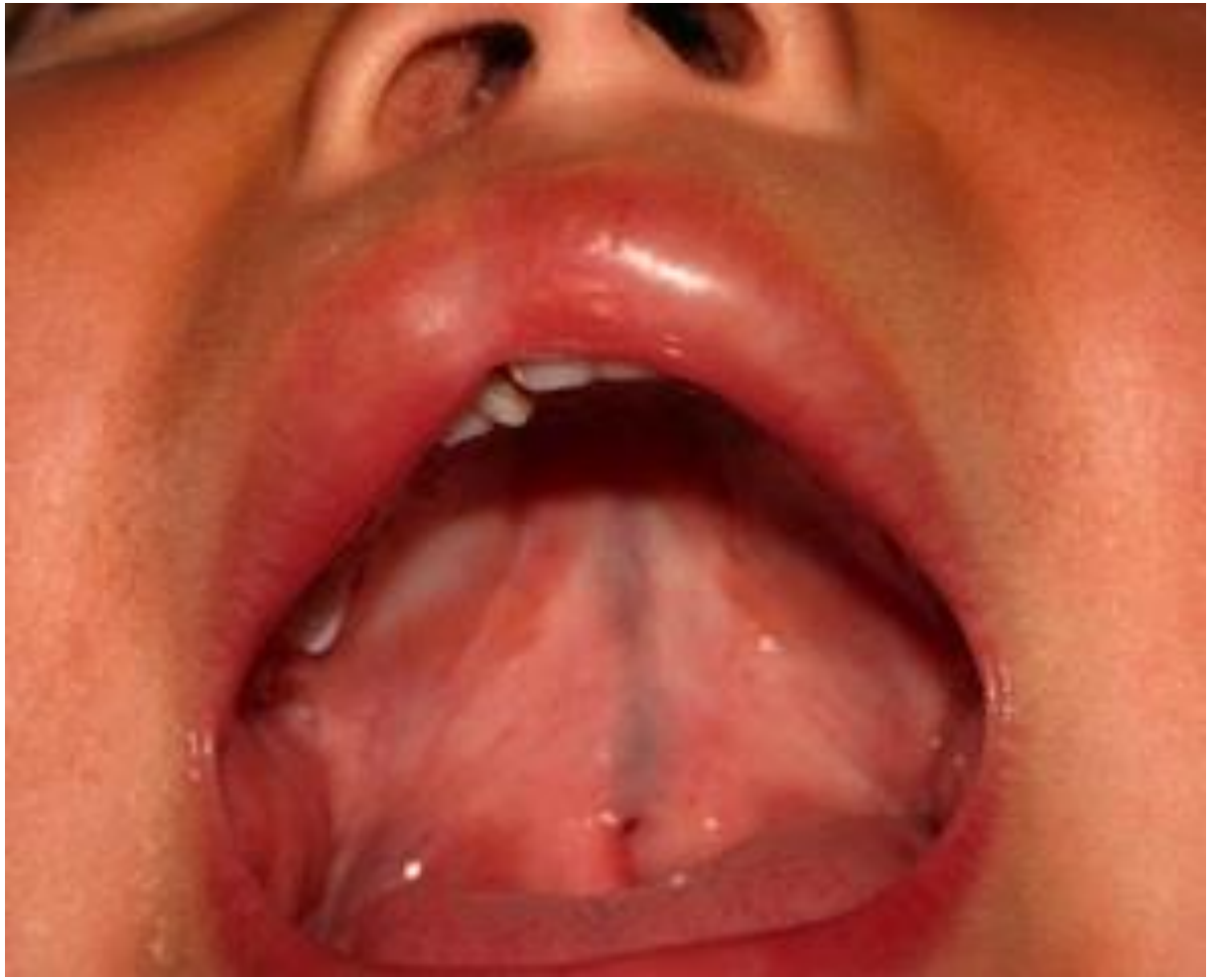
- Mandibular hypoplasia (Pierre-Robin syndrome)

  CAVE ! inspiratory airway obstruction

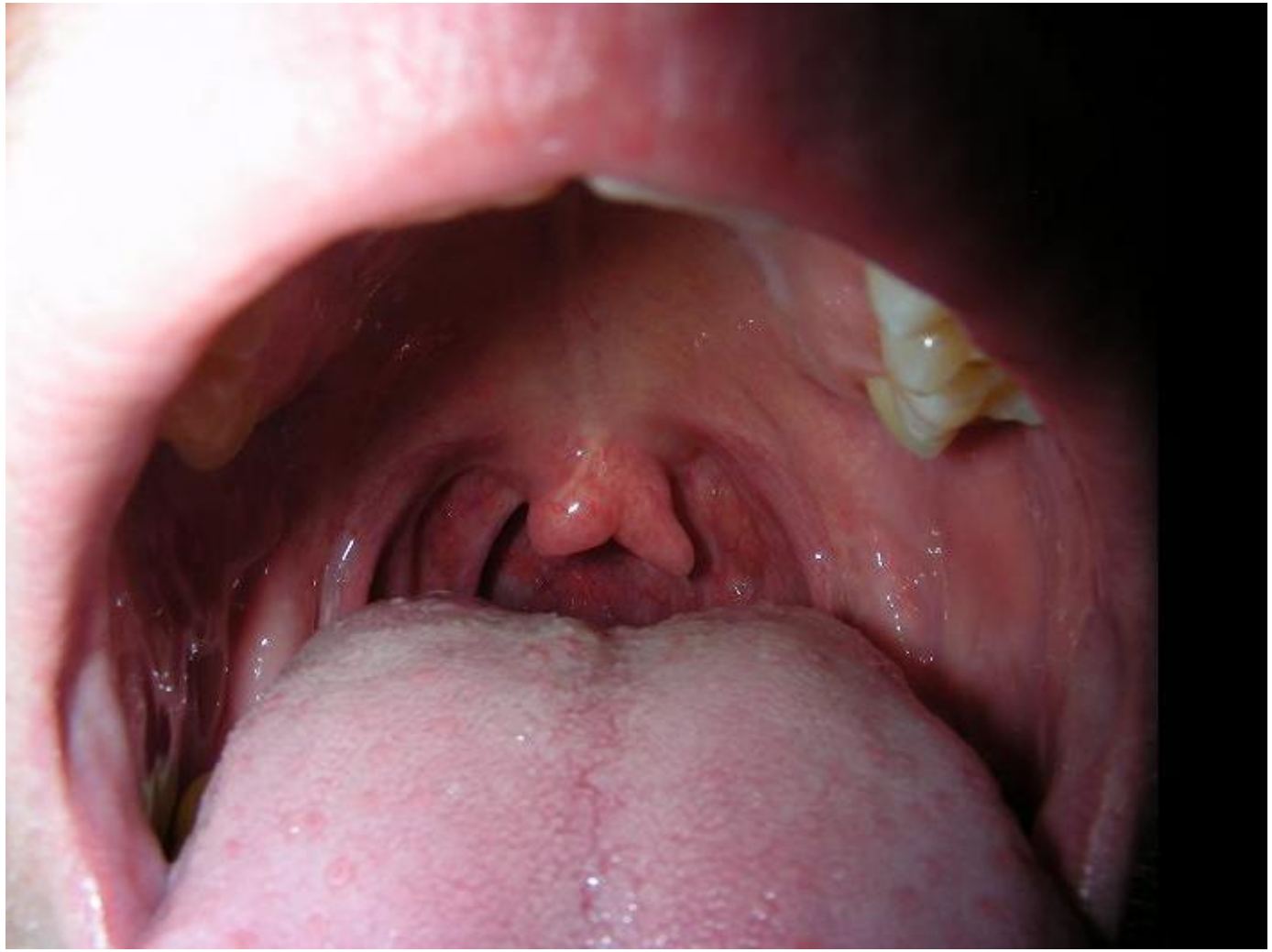
  - Micrognatia, retrognatia
  - High arched or cleft palate
  - Glossoptosis – with foreshortened floor of the mouth

http://emedicine.medscape.com/article/995706-overview

- Cleft lip and palate
- High arched palate – common association with limited nasal breathing



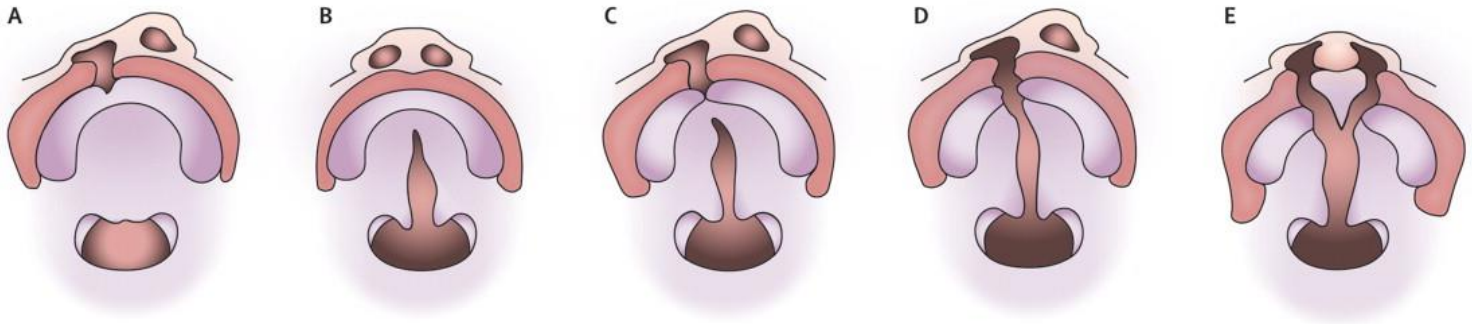
http://bestpractice.bmj.com/best-practice/monograph/675/resources/image/bp/9.html



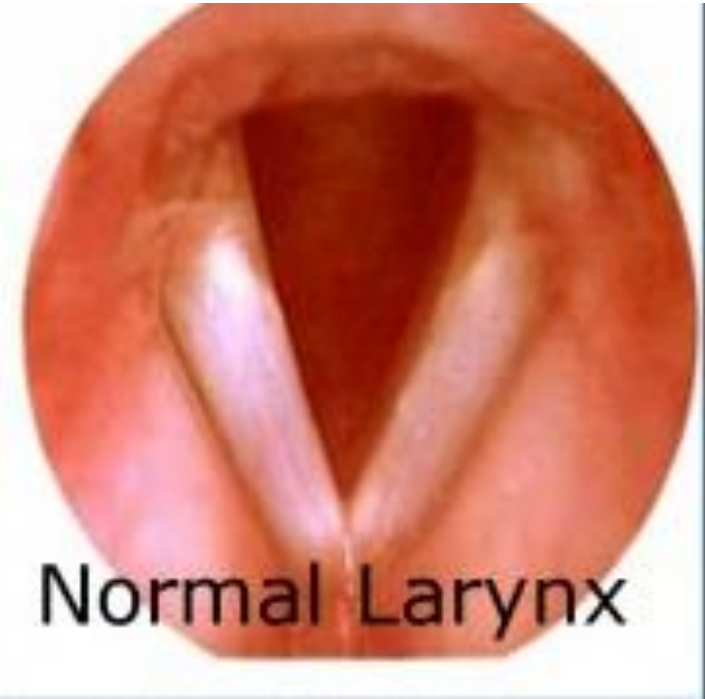
http://en.wikipedia.org/wiki/File:Bifid_uvula.JPG

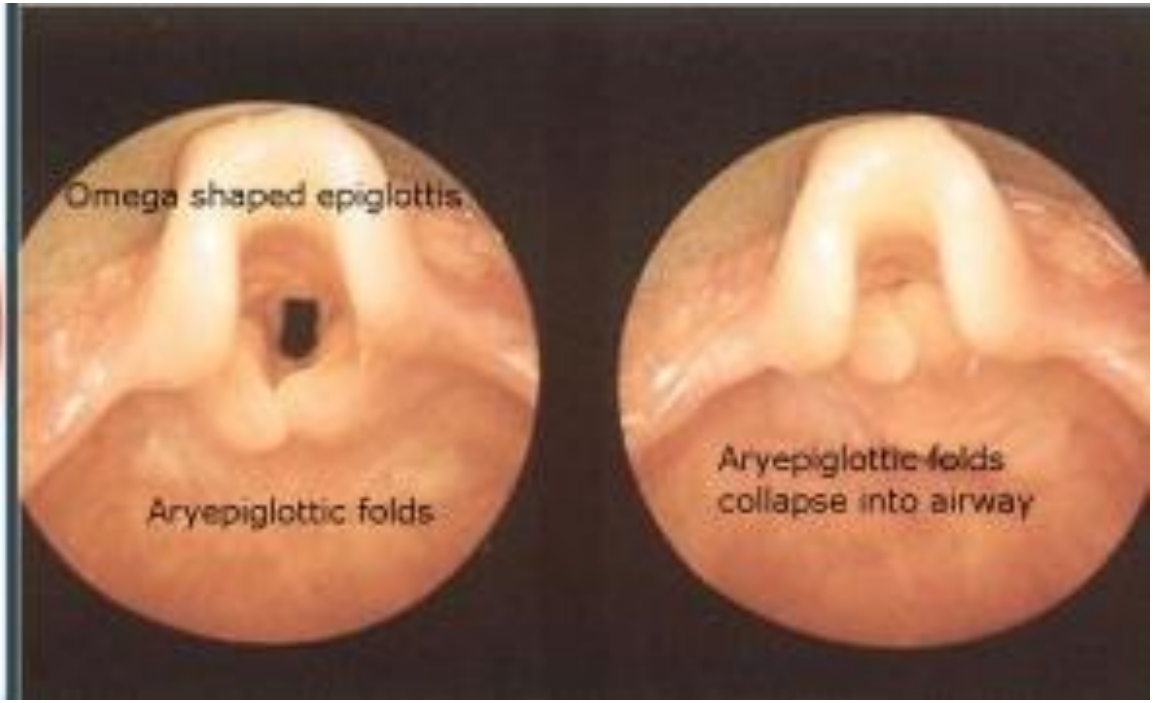
CLEFT LIP and PALATE

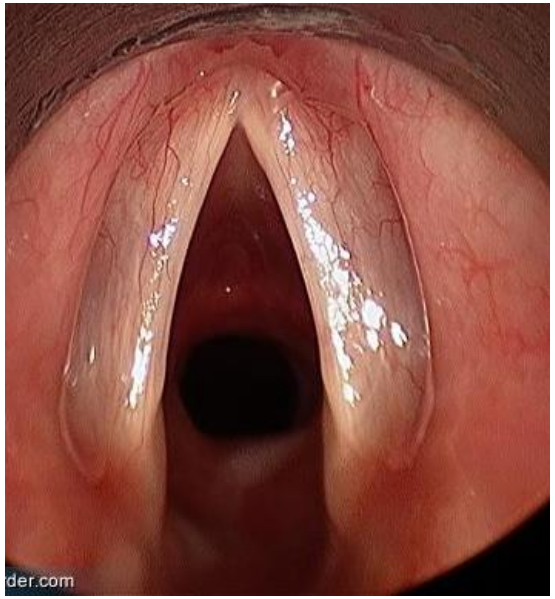
- Typical – cleft lip, lip and palate, isolated palate
- Atypical – facial – oblique, medial, lateral
  - Incidence - 1:750 – 2500, boys > girls
  - Sporadically > possible association with 1 of 400 syndrome
- Cleft unilateral x bilateral
- Manifestation variable from small notch in the uvula to complete separation



- http://linkinghub.elsevier.com/retrieve/pii/S0140673609606954?via=sd


Normal Larynx


Omega shaped epiglottis
Aryepiglottic folds
Aryepiglottic folds
collapse into airway



Congenital anomalies of the Larynx

- Pharyngeal x Laryngeal obstruction

  obstruction worse during sleep worse with activity

- Manifestation – <u>inspiratory stridor:</u>

1. Laryngomalacia – inspiratory stridor worse during crying or activity (feeding)

   usually appear within first 2 weeks

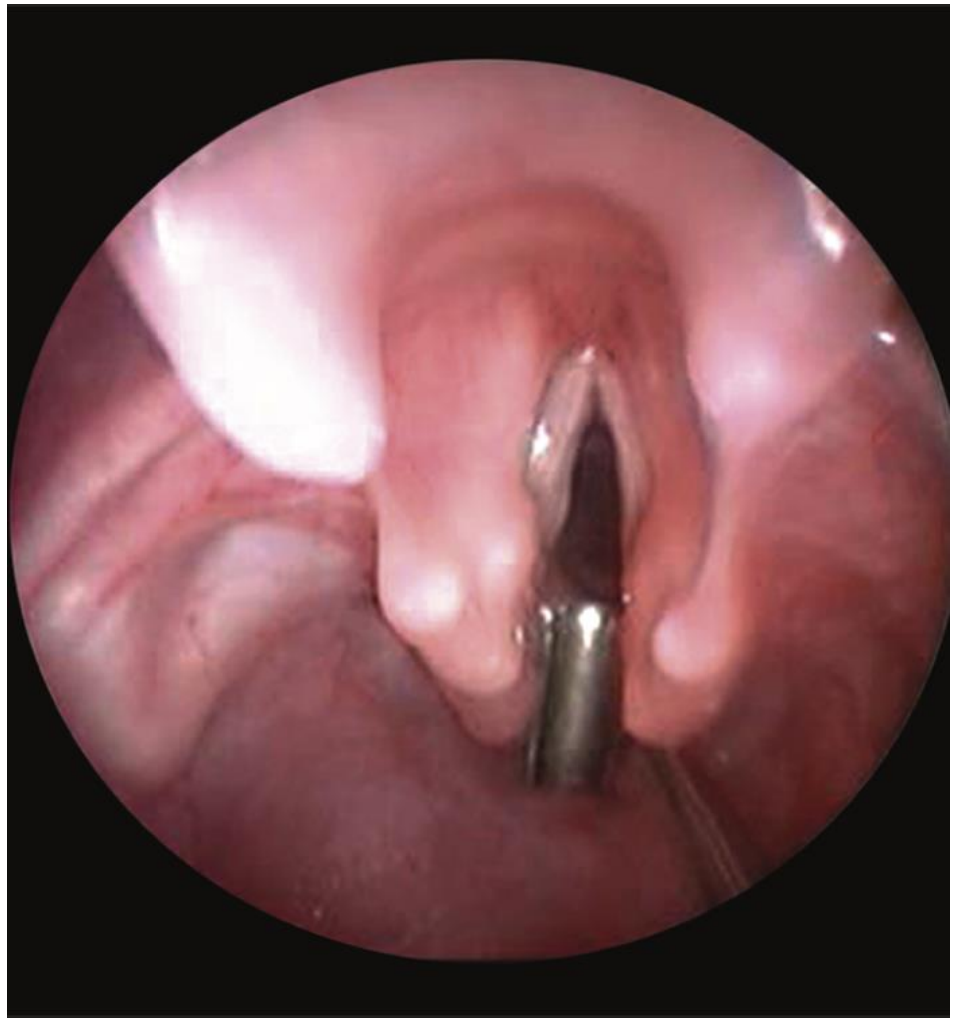
   15-60 % synchronous airways anomalies – complete bronchscopy in case of moderate to severe obstruction

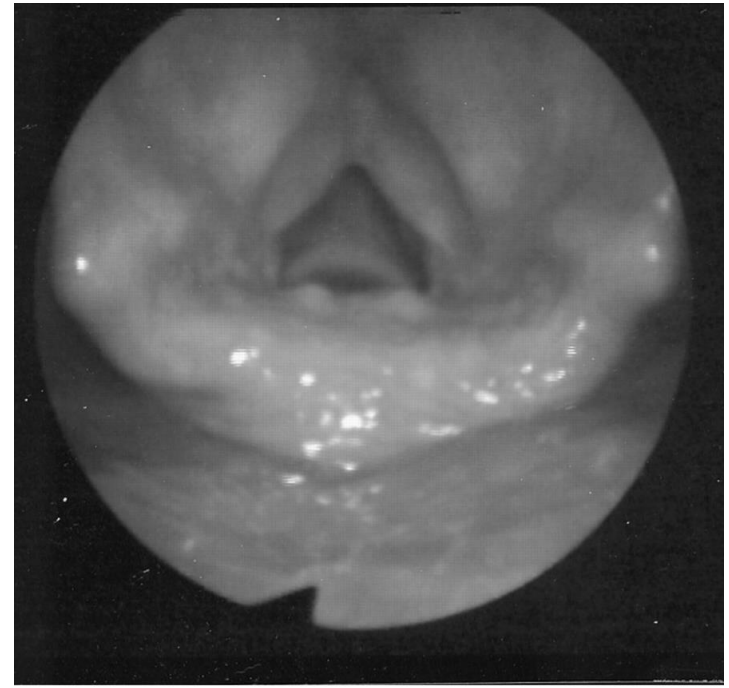
   common gradual improvement

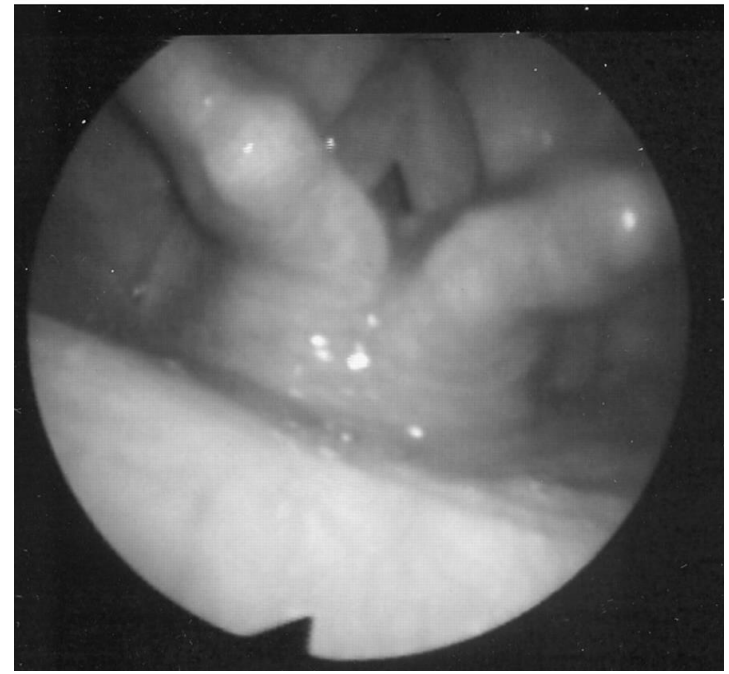
2. Congenital subglottic stenosis

   – recurent or persistent croup

   - usually cartilaginous







Congenital anomalies of the Larynx

Vocal Cord Paralysis- obvykle dobrá prognóza

Unilateral – aspiration, coughing and choking, wek and breathy crying

Bilateral – airway obstruction - stridor

Congental laryngeal web – glottic with subglottic extension = subglottic stenosis

Congenital subglottic hemangioma – hoarseness, stridor, barking cough

Posterior laryngeal cleft– symptoms of aspiration

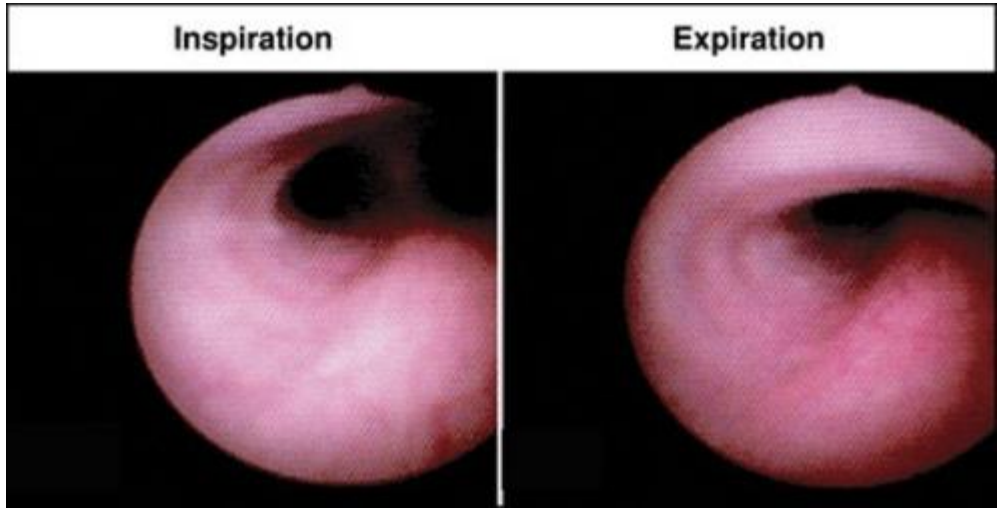
laryngotracheoesophageal cleft

Congential tracheal and bronchial anomalies

- Vascular and cardiac anomalies:

  Vascular ring or sling

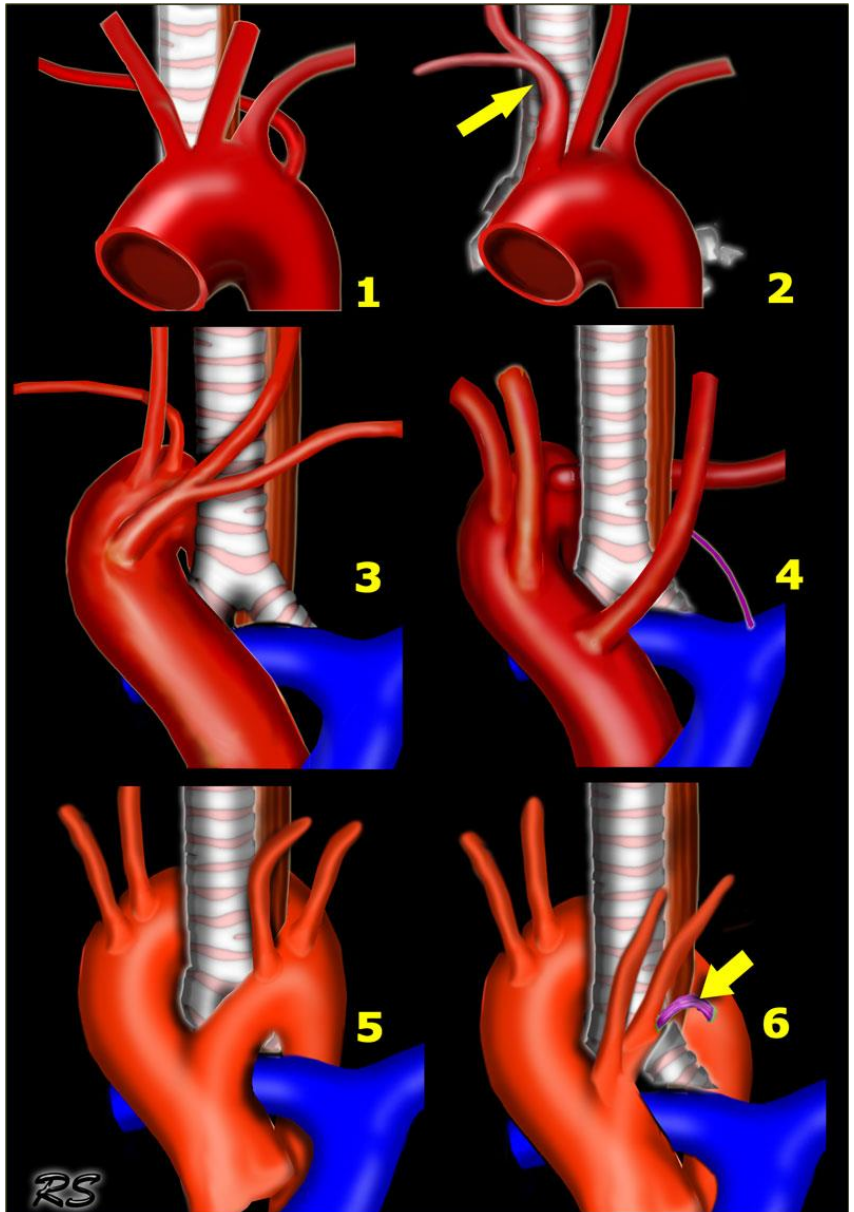
  = coughing, stridor, dyspnoea
- Tracheal stenoses, webs and atresia
- Tracheomalacia



Vicencio AG, Parikh S. In brief: laryngomalacia and tracheomalacia: common dynamic airway lesions. *Pediatr Rev*. 2006;27:e33-e35

VASCULAR ANOMALIES

1. Atyp. course of right artery
2. Sy a. innominata – truncus on the left
3. Right aortic arch
4. - „ – with left subclavian
5. Double arch
6. Right arch with ligamentum

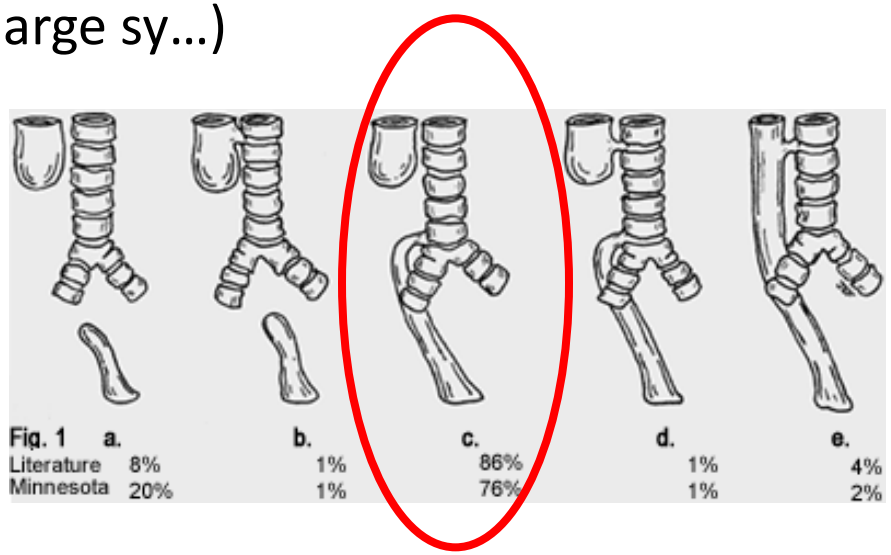


TRACHEOESOPHAGEAL FISTULA

- 1:3500, common assoc. with esophageal atresia
- 84 % trachea connected with distal esophagus
- 50% syndromic, other anomalies (Charge sy…)

Distal fistula

- early after birth - frothing
- cough, cyanosis,
- stomach distension
- aspirations



H- type fistula 4 %

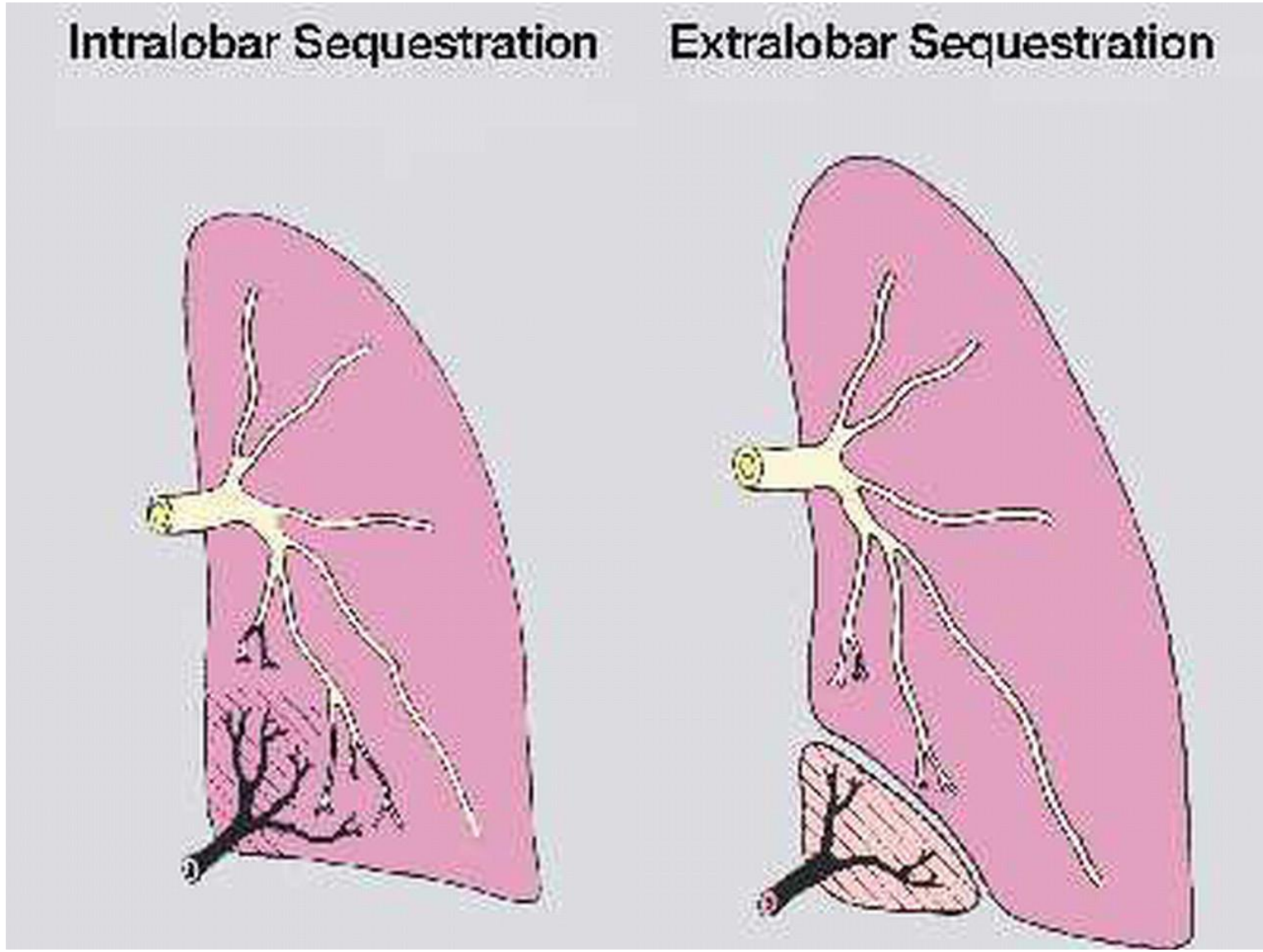
- later onset
- chronic respiratory problems (chronic bronchitis, pneumonias)
- resp. obtíže při jídle

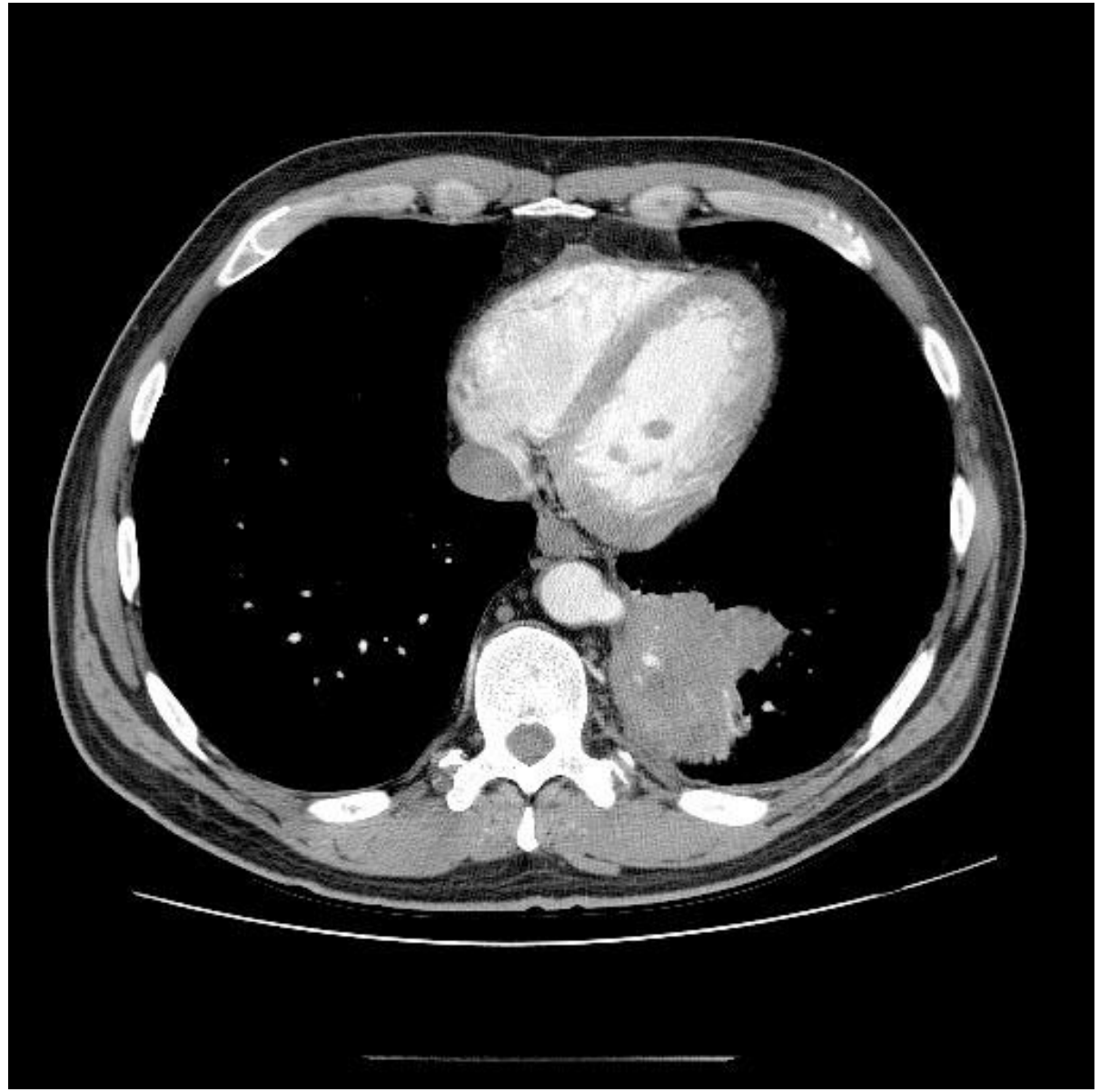
http://www1.umn.edu/eatef/whatis.html

Congenital disorders of the lung

- Pulmonary agenesis x pulmonary aplasia
- Unilateral left x right
- Pulmonary hypoplasia – limited space in thorax – limited breathing movements and/or ↓pressure of amnial fluidtlak amniál. tekutiny
  - Cystic adenomatoid malformation
  - Diaphragmatic hernia
  - Oligohydramnion – maternal disease, congenital renal anomaly
  - Congenital neuromuscular disease

  Lower number of alveoli and airway generations
- Pulmonary sequestration – extrapulmonary or intrapulmonary
  - Lung tissue withou connection with bronchus, arterial supply from the systemic arteries
  - Repeated infections, expansion


Intralobar Sequestration
Extralobar Sequestration



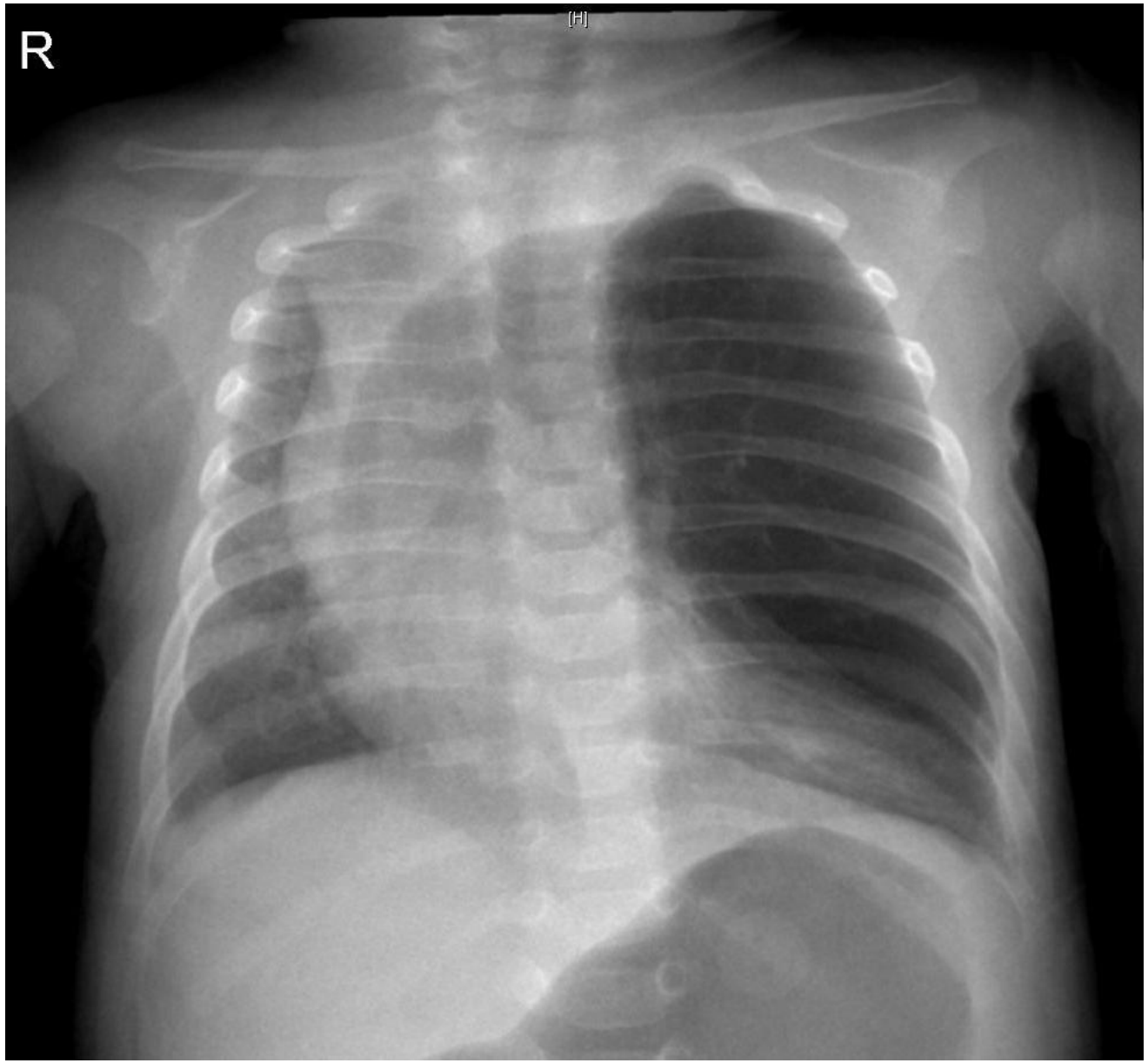
Congential disease of the lungs

Congenital lobar emphysema – neonatal period – respiratory distress, congenital overdistension of affected lobe – shift of mediastinus, atelectasis of normal lung tissue

- Immediative surgery x conservative treatment

Cystic adenomatoid malformation – cystic dysplastic lung tissue of one lobe – different types with variable prognosis

- Respiratory distress in early infancy
- Recurrent pneumonia, pneumothorax
- Surgery for symptomatic patients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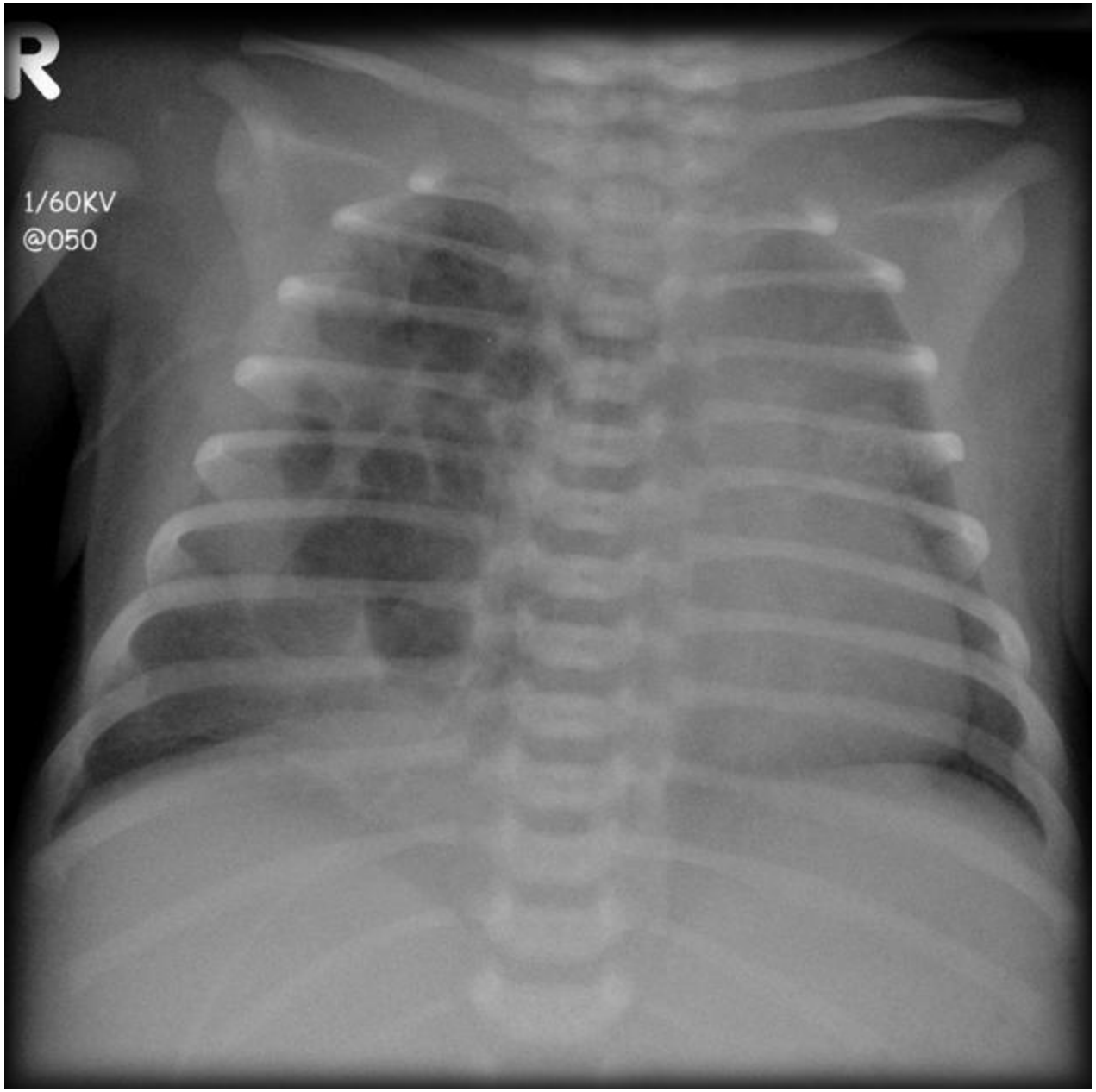
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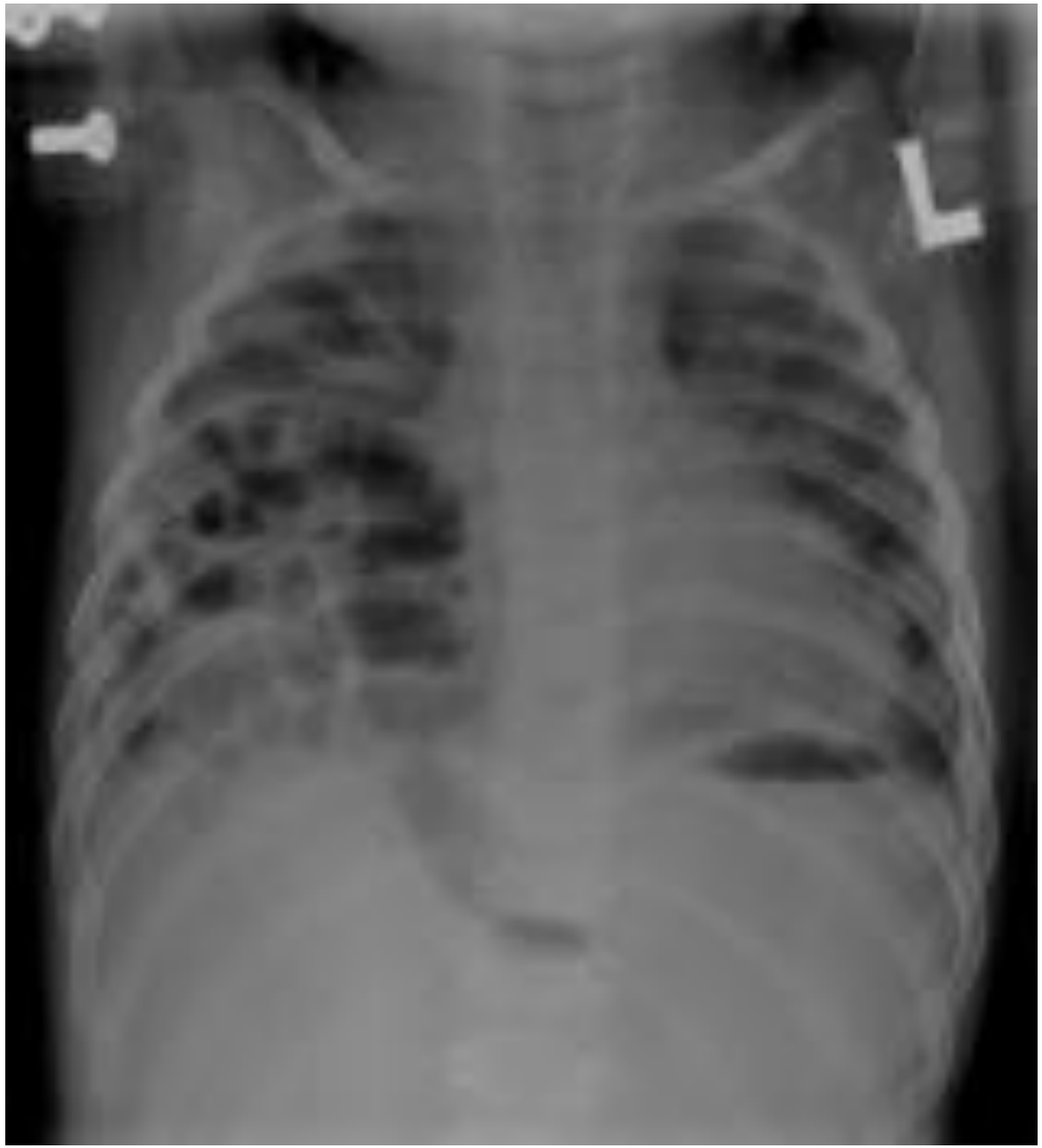
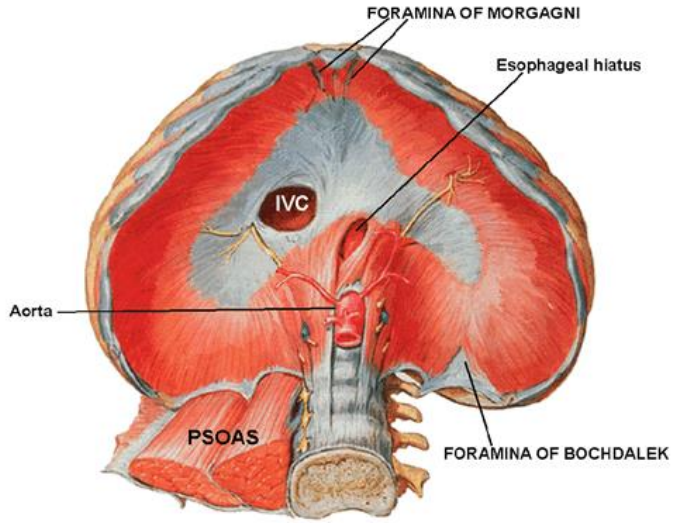

L

CONGENITAL DIAPHRAGMATIC HERNIA

- 1:2000-1:5000
- Pulmanory hypoplasia, pulmonary hypertension
- Prenatal diagnosis
- Early respiratory distress of neonates
- Manifestation – weak breathing sounds, niveau of abdominal wall bellow – scaphoid abdomen, shift of heart sounds (mediastinum) bowel sounds in the chest

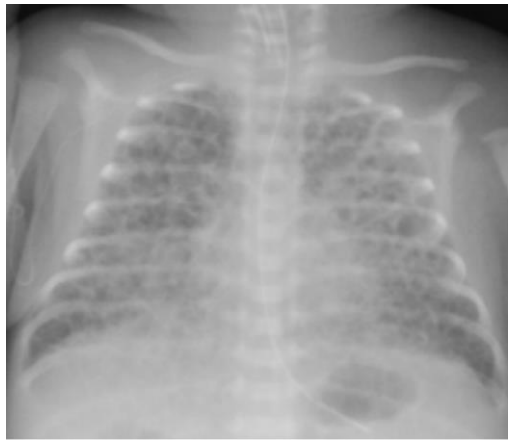
X- RAY

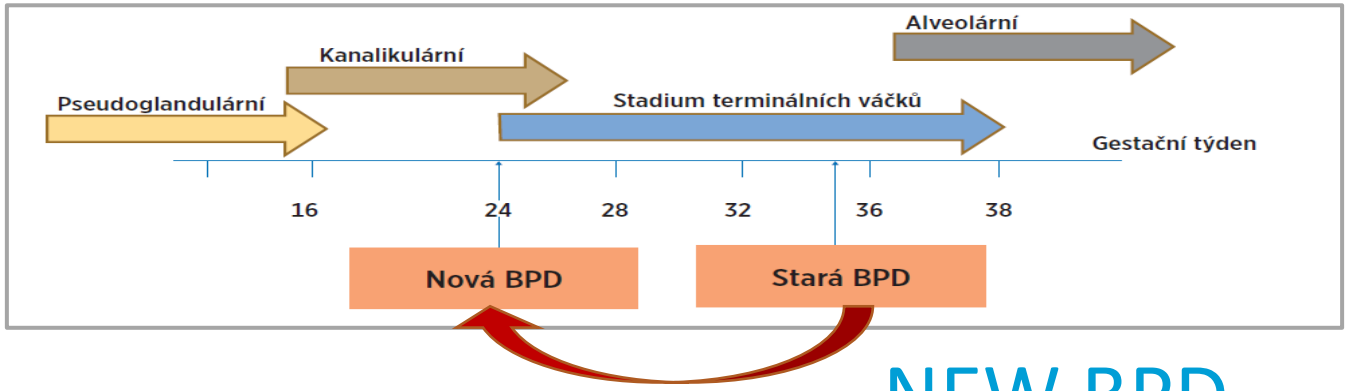
- transport in utero – specialized centrum
- Orotracheal intubation + ventilatory support
  avoid resuscitation with ambuvac with mask
- Nasogastric tube - stomach air bubble

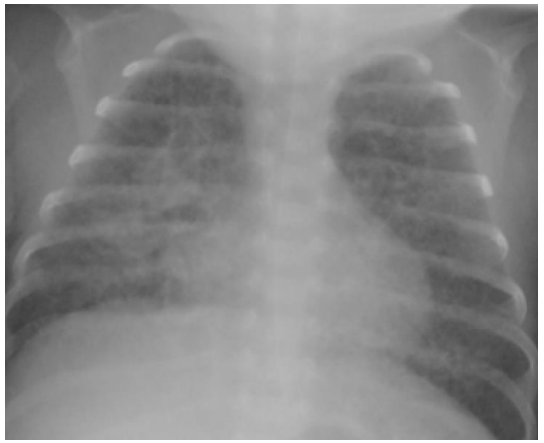


x

BRONCHOPULMONARY DYSPLASIA







OLD BPD

FIBROPROLIFERATION

- Arteficial lung ventilation
- Hign concentration of 0_2
- High mortality
- Limited survival below 28. gw

Northway 1967 - 31. x 34 gt

NEW BPD

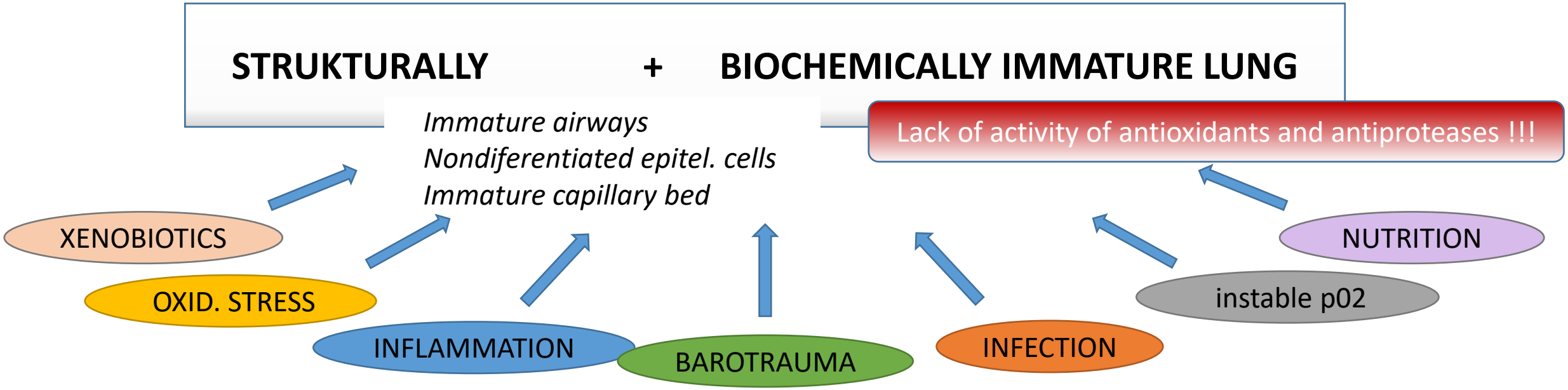
FAILURE OF DEVELOPMENT

- Antenatal steroids
- Surphactant
- Noninvasive respiratory support = ↓ 0_2
- Persistent ductus arteriosus - therapy
- Nutrition

Jobe et al. 1999

Even in children with minimal noninvasive respiratory support !!!!!

PREMATURITY x RESPIRATORY SYSTEM



- Rapid change of mechanical properties (fetal breathing movements, amniotic fluid circulation) ↔ spontaneous or artificial ventilation
- Physiologically low values of $p0_2$ in utero ↔ even room air with 21 % is relatively hyperoxic