بسم الله الرحمن الرحیم

بیماریهای پلور در نوزادان

دکتر صدرالدین مهدی پور

Chylothorax

- Chylothorax is the accumulation of lymphatic fluid (chyle) in the pleural cavity.
- It is a rare entity, with a reported incidence of 1 :10,000 births, and affects males more than females (2:1)
- It is the most common cause of pleural effusion in neonates and can be primary (congenital) or secondary (acquired).

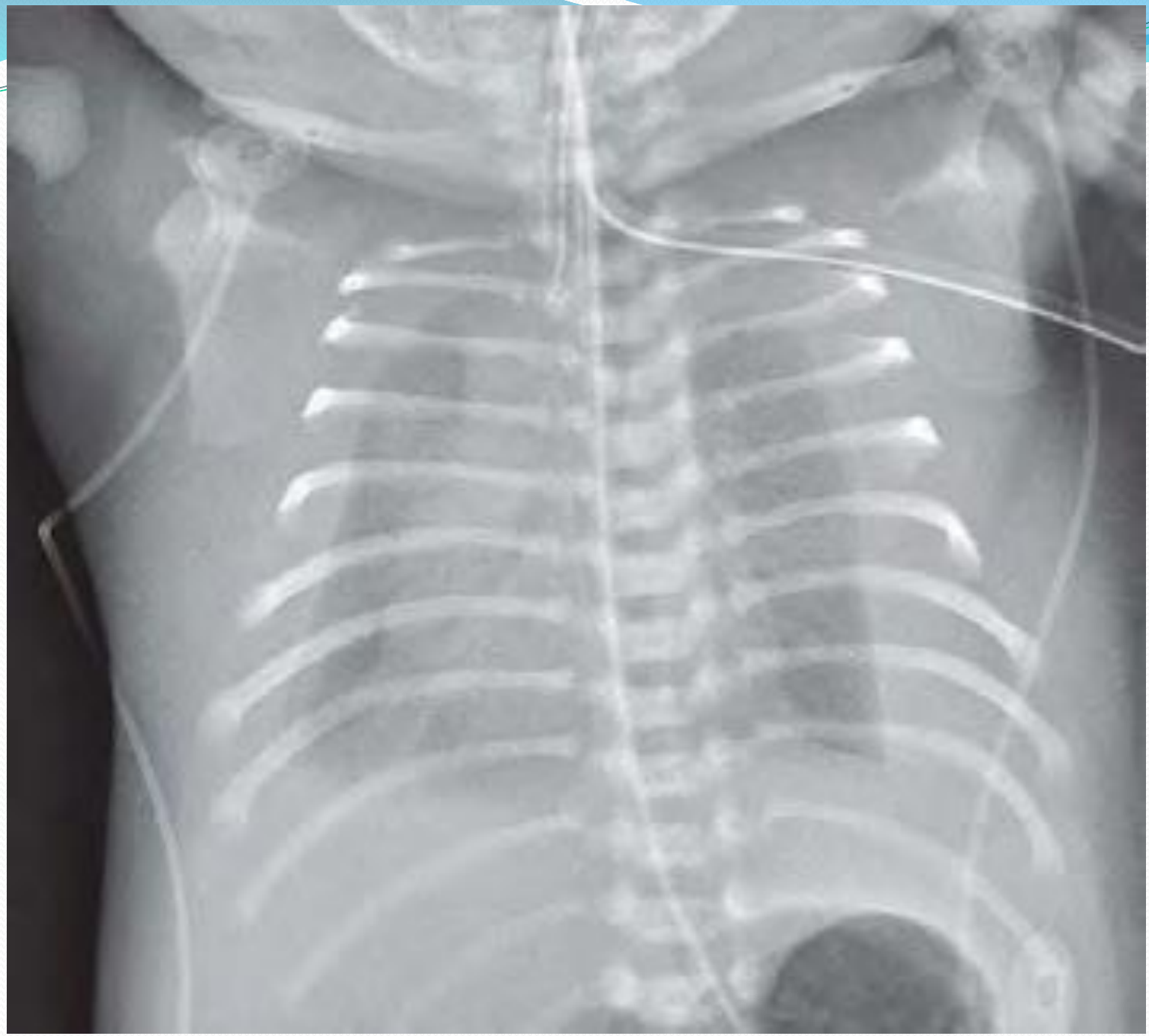
- Congenital chylothorax(less than 10% of all chylothoraces) may be associated with abnormalities of the lymphatic system.
  - Congenital heart disease
  - mediastinal malignancies
  - chromosomal abnormalities such as trisomy 21, Noonan syndrome, or Turner syndrome.

- Secondary chylothoraces are most commonly associated with trauma during thoracic surgery or result of increased superior vena caval pressure caused by **venous thrombosis.**

- Clinical presentation is that of respiratory distress secondary to **lung compression**, **pulmonary hypoplasia**, or symptoms of the **underlying pulmonary or cardiac disease.**

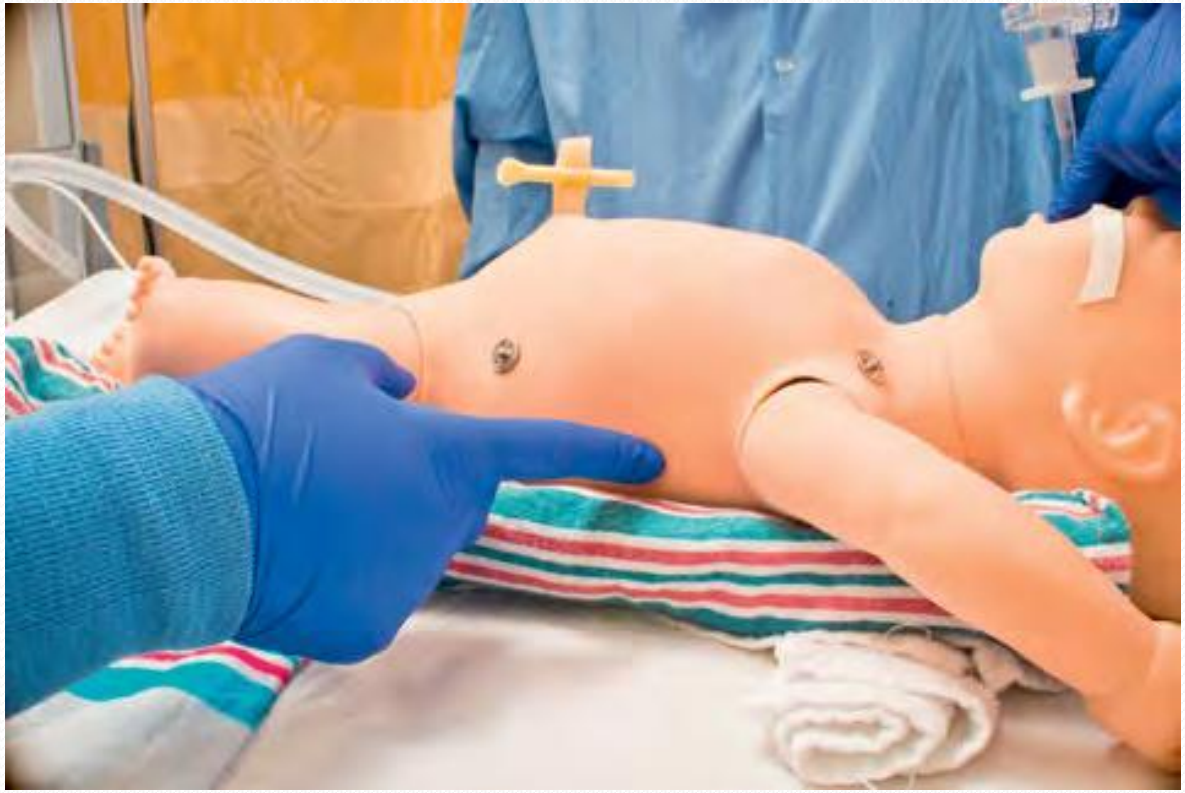
- Diagnosis of chylothorax are by **prenatal ultrasound** (antenatal management consists of thoracocentesis or thoracoamniotic shunt placement) to try to prevent the development

- **Physical examination** is significant for decreased breath sounds on the affected side with shifting of the cardiac apex to the contralateral side.
- **Chest radiograph** shows a pleural effusion, compression of the lung on the affected side, and displacement of the heart to the opposite side.



- Diagnosis is established by analysis of the pleural fluid. In neonates with established feedings, chylothorax appears **milky in color**; however, in nonfed neonates, it is **clear**.
- Buttiker and colleagues have proposed the following criteria for establishing the diagnosis of chylothorax: **absolute cell count of greater than 1000/µL with a lymphocyte fraction of greater than 80% and triglyceride levels greater than 1.1 mmol/L (97.43mg/dl)**

For a pleural effusion, the aspiration site is the fifth or sixth intercostal space along the posterior axillary line. Place the baby on his back (supine) to allow the fluid to collect in the lower (posterior) portion of the chest



- Optimal treatment for chylothoraces has not been defined but is **mostly supportive** while awaiting resolution of the effusion.
- Mechanical ventilation and drainage of the chylothorax might be needed in patients with large effusions, and nutritional support using total parenteral nutrition is essential.
- When feedings are started, formulas containing a high percentage of medium-chain triglycerides (MCTs) are recommended, **because lymphatics are not needed for MCT absorption.**
- **In most cases, spontaneous resolution occurs within 4-6 weeks.**

Table 1. Comparison of Monogen and P

Composition Per Liter	Monogen
Energy (kcal)	740
Fat (g)	20
% MCT	93
LCT per Litre (g)	1.4
Protein	Whey protein concentrate and amino acids
Osmolality at standard dilution (mosm/kg)	280
Essential fatty acids provided by	Walnut oil

LCT = long-chain triglyceride; MCT = medium-chain triglyceride.

- Several treatment strategies have been described for cases with persistent chylothorax, including **pleurodesis, ligation of the thoracic duct**, and **pleuroperitoneal shunt.**
- Whereas povidone-iodine pleurodesis has been used successfully in persistent chylothorax, it has also been associated with renal failure.
- There is growing evidence from uncontrolled case studies suggesting a markedly positive effect of somatostatins, particularly octreotide, in the treatment of chylothorax with minimal side effects.
- **In the absence of a controlled trial evaluating safety and efficacy, this therapy should be reserved for persistent and severe cases and not as first line of treatment.**

Congenital Pulmonary Lymphangiectasia

- characterized by dilation of lymphatic vessels in multiple areas of the lungs.
- first described by Virchow in 1856
- Most cases of CPL are sporadic with a predilection for male involvement (2:1)

- The characteristic pathologic finding of CPL is pulmonary lymphatic dilation in the subpleural, interlobar, perivascular, and peribronchial lymphatics.
- It may be associated with nonimmune hydrops fetalis and congenital chylothorax.

- CPL is classified as primary or secondary.
- Primary CPL can present as either a primary pulmonary developmental defect that can be localized or diffuse or as a part of a more generalized lymphatic developmental defect.
- Patients with generalized lymphangiectasia tend to have less severe pulmonary involvement.

- Secondary cases of CPL are often associated with **cardiac malformations with obstructed pulmonary venous return including:**
  - **obstructed total anomalous pulmonary venous return**
  - **hypoplastic left heart syndrome**
  - **cor triatriatum**(the left atrium is divided into two chambers with an opening)

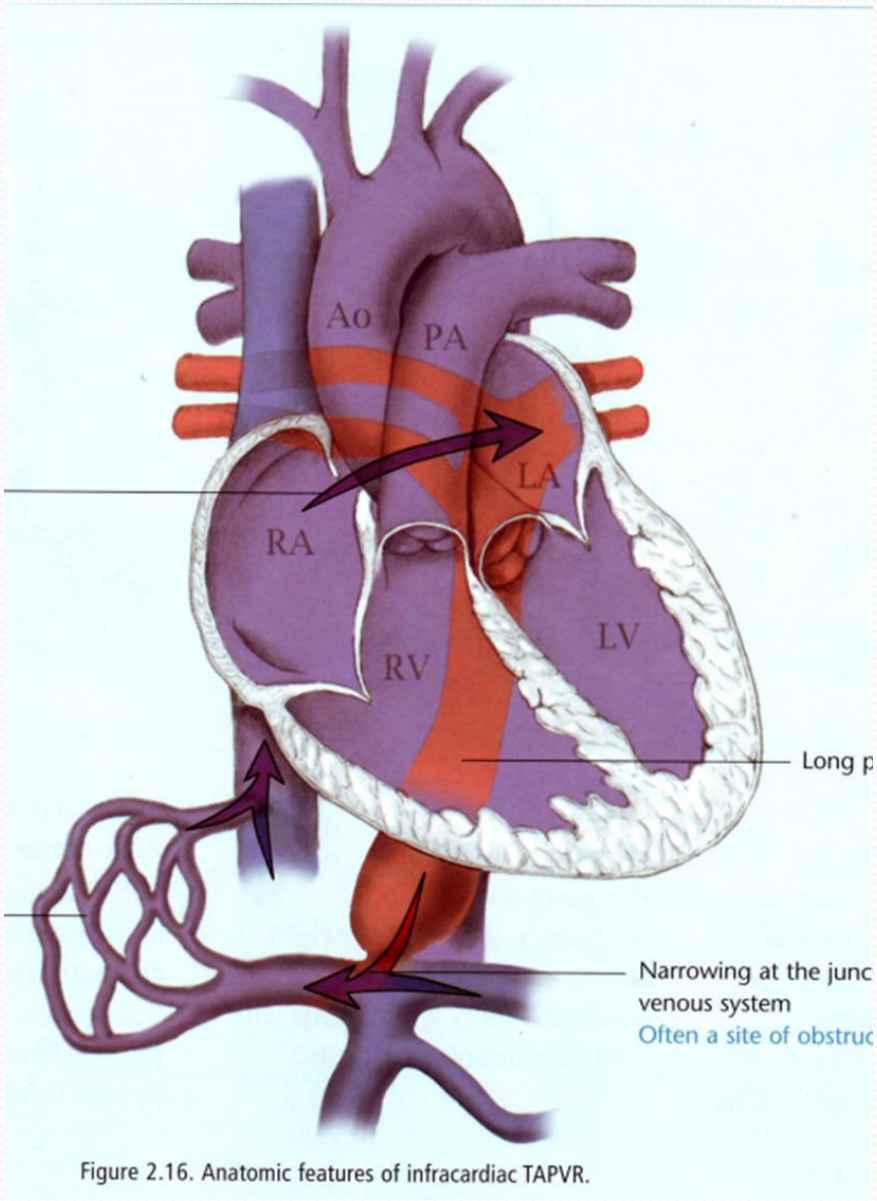
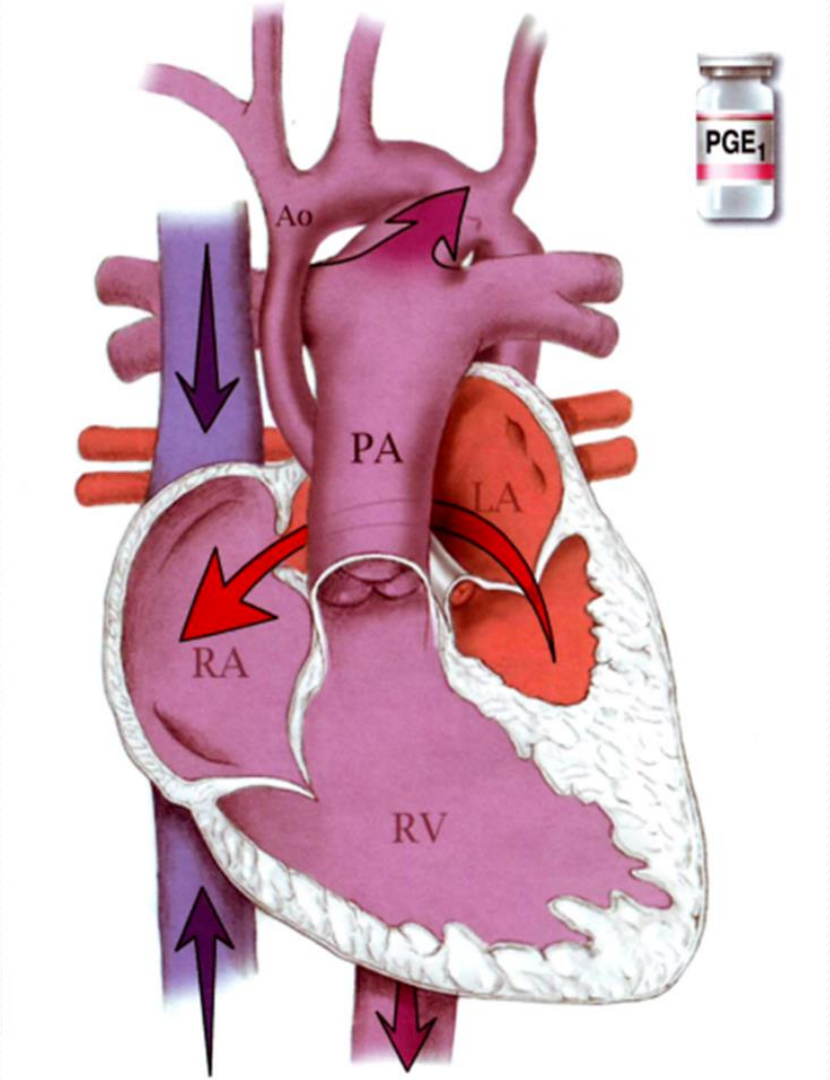
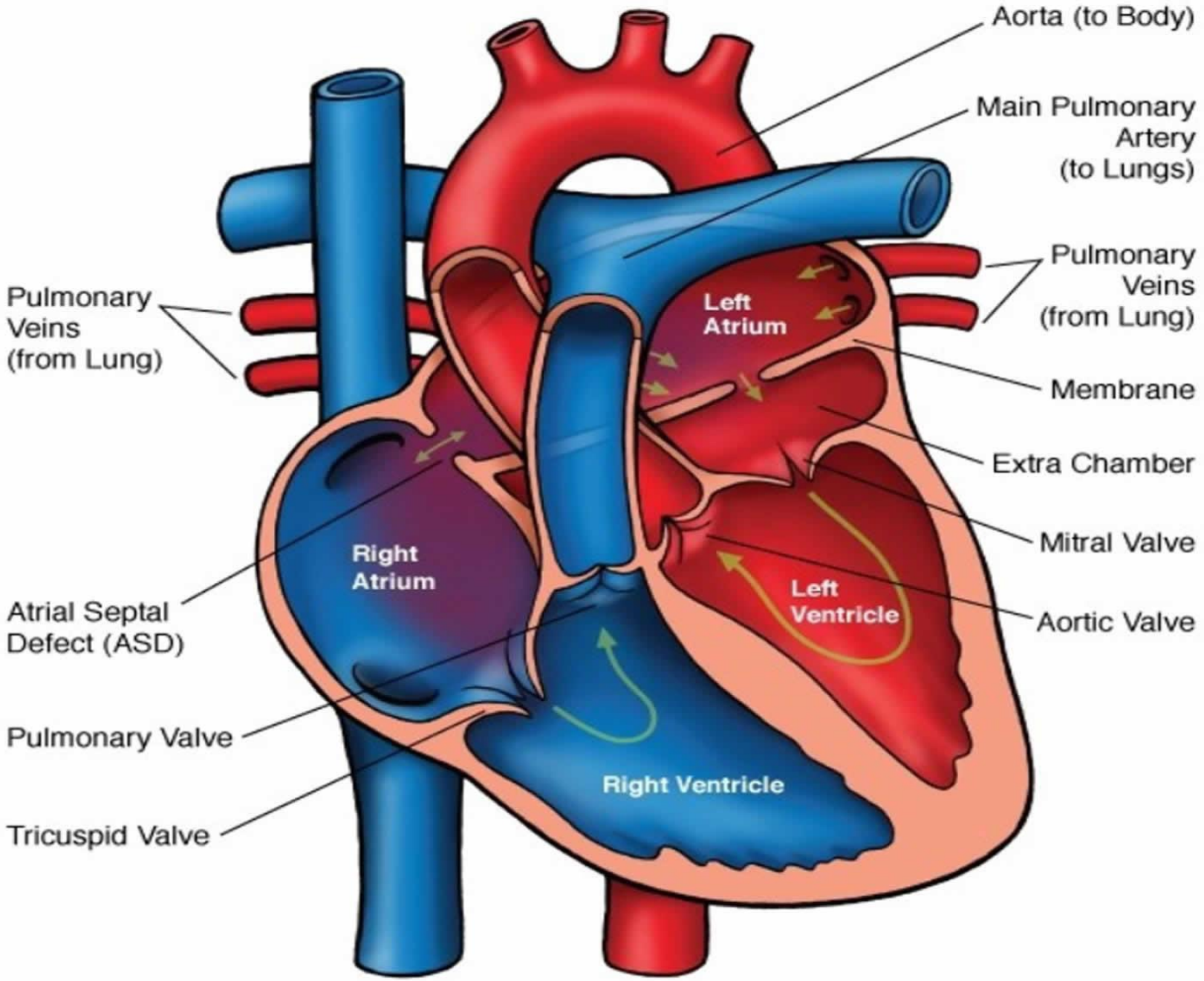


Figure 2.16. Anatomic features of infracardiac TAPVR.



through the ductus arteriosus to perfuse the body, including the head, neck and coronary vessels.

Cor Triatriatum



- Congenital pulmonary lymphangiectasia has also been described in multiple syndromes, including Noonan, Down, and Ullrich-Turner

- Patients with CPL usually present with **intractable respiratory failure**, **cyanosis**, and **hypoxia associated with bilateral chylothoraces in the first few hours of life**, although diagnosis can be delayed for several weeks in cases of unilobar involvement.

- Examination of the pleural fluid shows characteristic findings of chylothorax including a lymphocytosis and elevated triglycerides, although elevated triglycerides might be absent in nonfed infants

- **Chest radiograph** reveals hyperinflation of the lung with bilateral interstitial infiltrates and bilateral pleural effusions.
- **High-resolution computed tomography** demonstrates diffuse thickening of the peribronchovascular interstitium and the septa surrounding the lobules.

- **Definitive diagnosis** is made by **lung biopsy** showing the characteristic features: increased fibrous tissue with dilation of cystic lymphatic spaces and collapsed alveoli, although differentiation from lymphangiomatosis can be difficult.

- Treatment is mostly supportive. Intubation and mechanical ventilation; drainage of pleural and peritoneal effusions;and correction of hypoxia, acidosis, and shock might be needed in the delivery room for stabilization.
- Persistent chylothorax might require chest tube placement.

- Nutritional therapy with medium-chain triglycerides and total parenteral nutrition has been successful in the treatment of CPL.
- Pleurodesis with sclerosing agents has been used to treat persistent chylothoraces associated with the disease.
- More recently, reports of using lymphangiogram with ethiodized oil as a successful treatment modality have been reported.

- **The prognosis appears to depend on the severity of symptoms in the immediate newborn period.**
- Although traditionally thought to be fatal, there are reports of survival in some patients presenting with respiratory failure, chylothorax, and hydrops fetalis in the immediate neonatal period.
- Later presentation carries a better prognosis with the possibility of spontaneous resolution, although respiratory morbidity might be common.

Pulmonary Air Leak Syndromes

- Air leak occurs more commonly in the newborn period than any other period of life.
- Air leak syndromes include:
- pneumothorax,
- pneumomediastinum,
- pulmonary interstitial emphysema,
- pneumopericardium,
- pneumoperitoneum
- subcutaneous emphysema.

- Even though air leaks can occur spontaneously, they mostly occur in patients with **lung pathology**, including MAS, pneumonia, RDS, diaphragmatic hernia, and pulmonary hypoplasia, especially when positive-pressure ventilation is required.

- The introduction of surfactant for treatment of RDS has caused a decrease in the incidence of air leaks.
- Conversely, the increasing practice of elective c/s before 39 completed weeks' gestation is associated with increased incidence of pneumothorax when compared with emergent cesarean section or vaginal delivery.

Pathophysiology

- **Atelectasis** in RDS and **plugged small airways** in MAS cause unequal distribution of the ventilated volume and transpulmonary pressure to the more distensible areas of the lung, increasing the risk of rupture and air leak.
- Partial obstruction during MAS causes air trapping when inspired air is not completely evacuated during exhalation

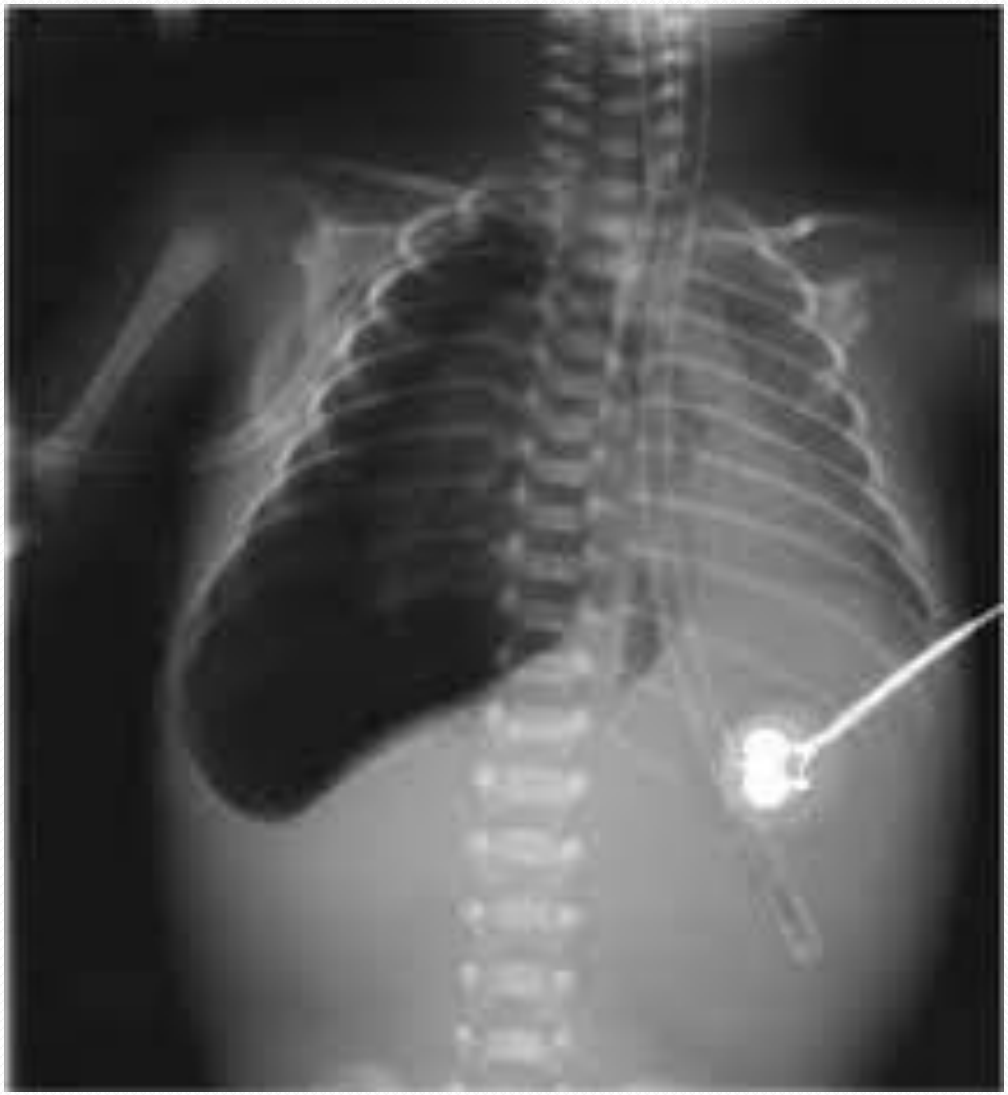
- Higher peak inspiratory pressure especially in the 24 hours preceding pneumothorax, **long inspiratory time** (>0.5 seconds), and **frequent suctioning in the 8 hours before pneumothorax.**

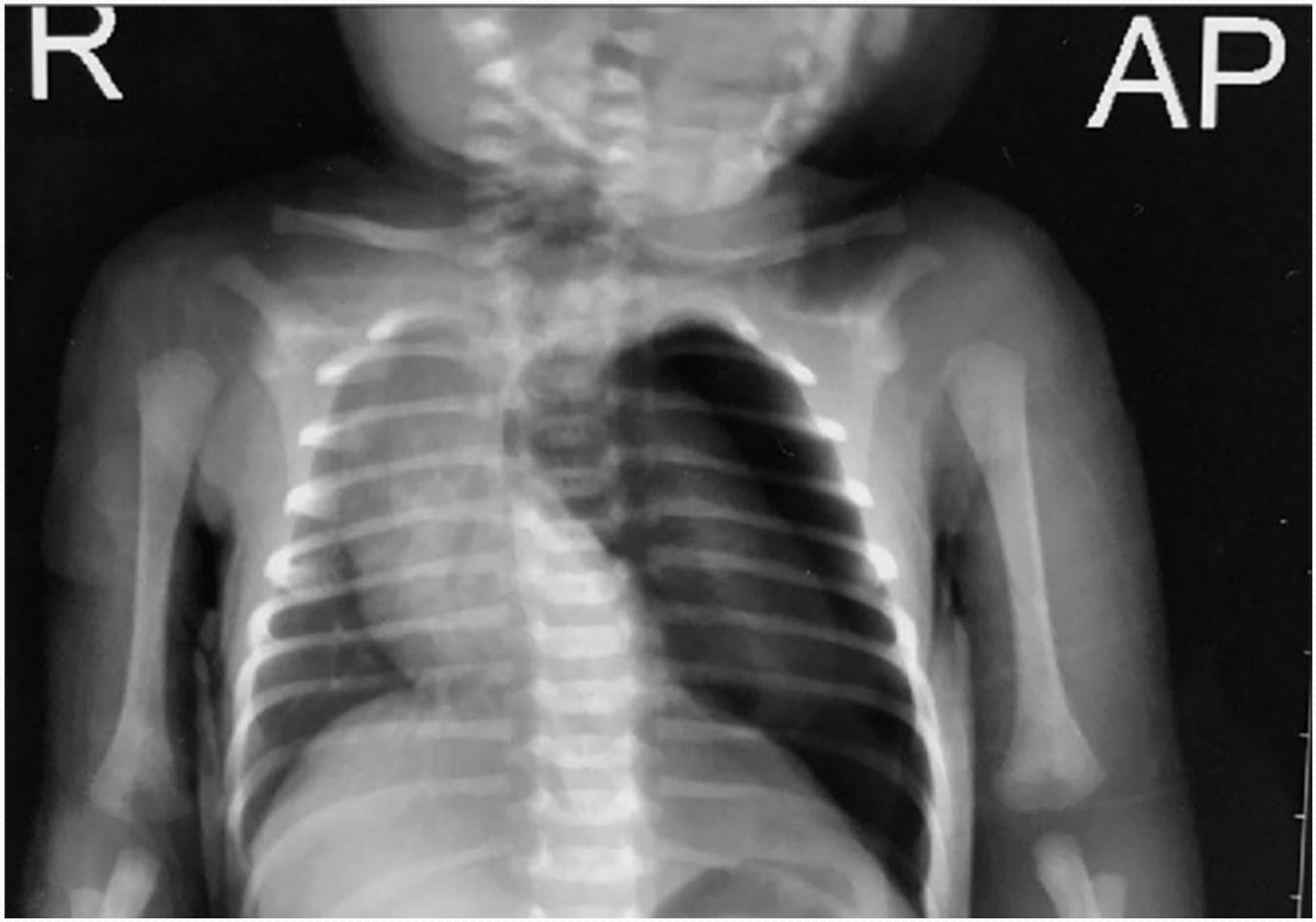
- Elective initial use of high nasal CPAP (8 cm H_2O) in infants at 25-28 weeks' gestation was associated with increased incidence of air leak from 3%-9% when compared with intubated infants who received surfactant therapy.

Clinical Presentation

- Sudden worsening in respiratory distress should always raise the suspicion of the development of a pneumothorax.
- Tachypnea, grunting, flaring, and retractions occur most commonly.
- Physical examination reveals **distant breath sounds**, **overdistension of the chest wall**, and **bulging abdomen** on the affected side secondary to downward displacement of the diaphragm in patients with unilateral pneumothorax.

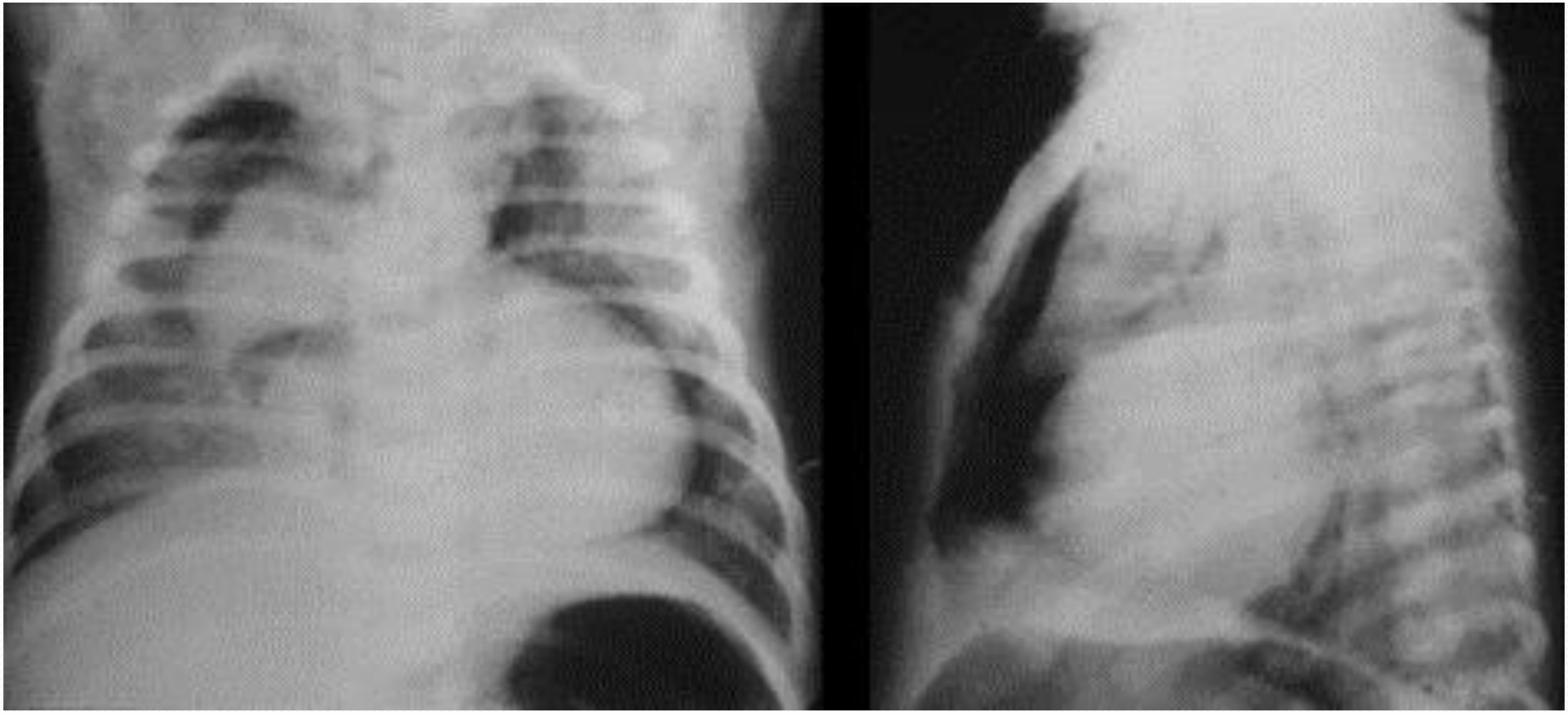
- **Larger volumes of leaked air** can cause significant increase in intrathoracic pressure, which impairs venous blood return and compromises cardiac output, causing poor tissue perfusion and metabolic acidosis.




R
AP

- Pneumomediastinum is often asymptomatic if not associated with pneumothorax and is mostly found on radiographic evaluation of patients with respiratory distress or distant heart sounds.
- Dissection of air through the anterior mediastinum into the neck can cause **subcutaneous emphysema** that is most commonly felt as subcutaneous crepitus in the face, neck, or supraclavicular notch area.

Air Leak Syndromes: Pneumomediastinum



Images from Virtual Children's Hospital

Pneumomediastinum

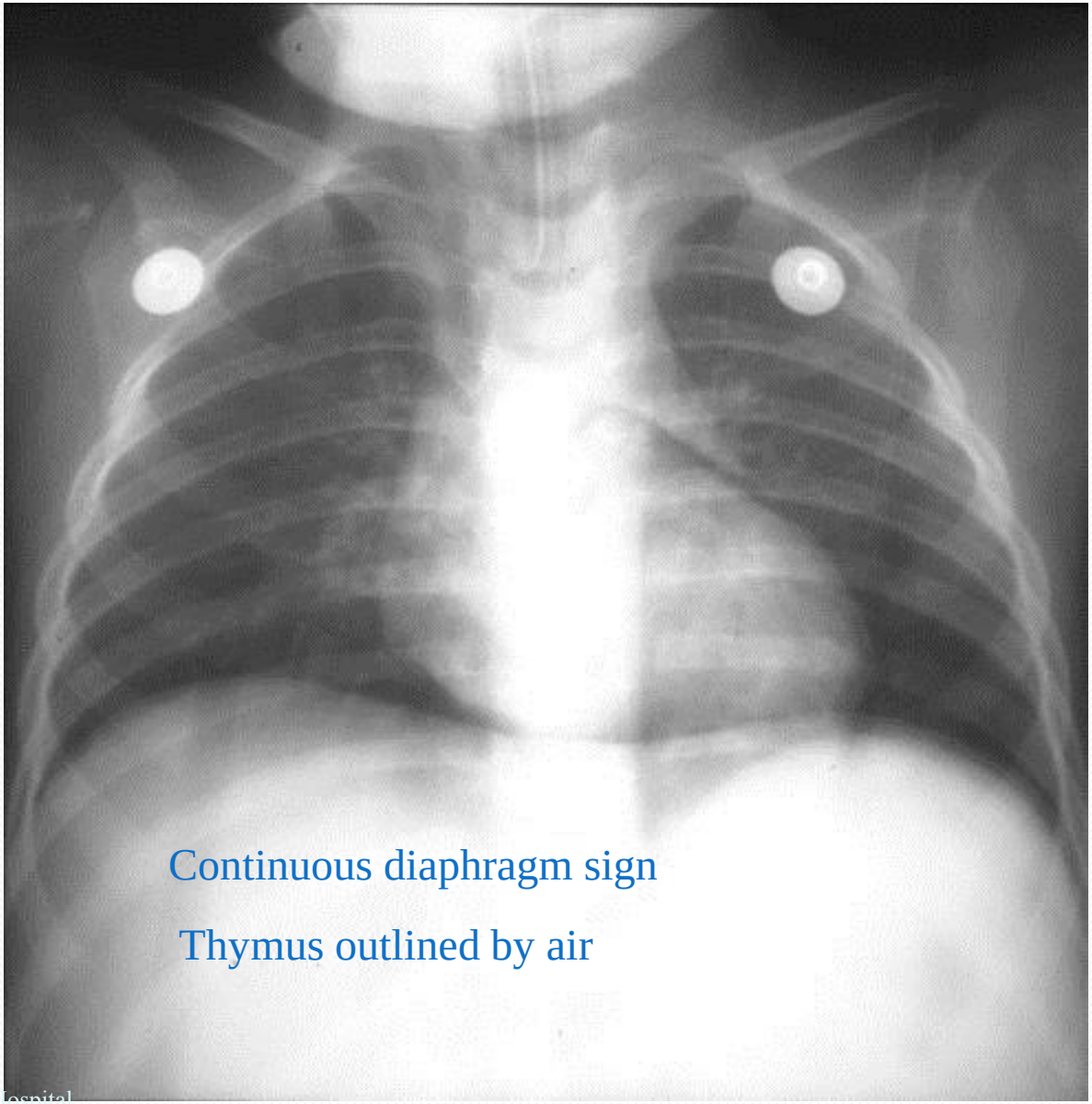


Image from Virtual Children's Hospital

- **Pneumopericardium** is a rare and serious complication probably caused by dissection of leaked air through the vascular bundle of great vessels.
- Trapped air in the limited pericardial space can quickly cause cardiac tamponade, **decreasing venous return**, and **cardiac output**.
- The clinical picture consists of worsening respiratory distress, hypotension, bradycardia, pallor, and/or cyanosis.
- Cardiac sounds are distant or muffled on auscultation and a pericardial rub might also be heard. Low voltage QRS complexes can also be seen.

Pneumopericardium

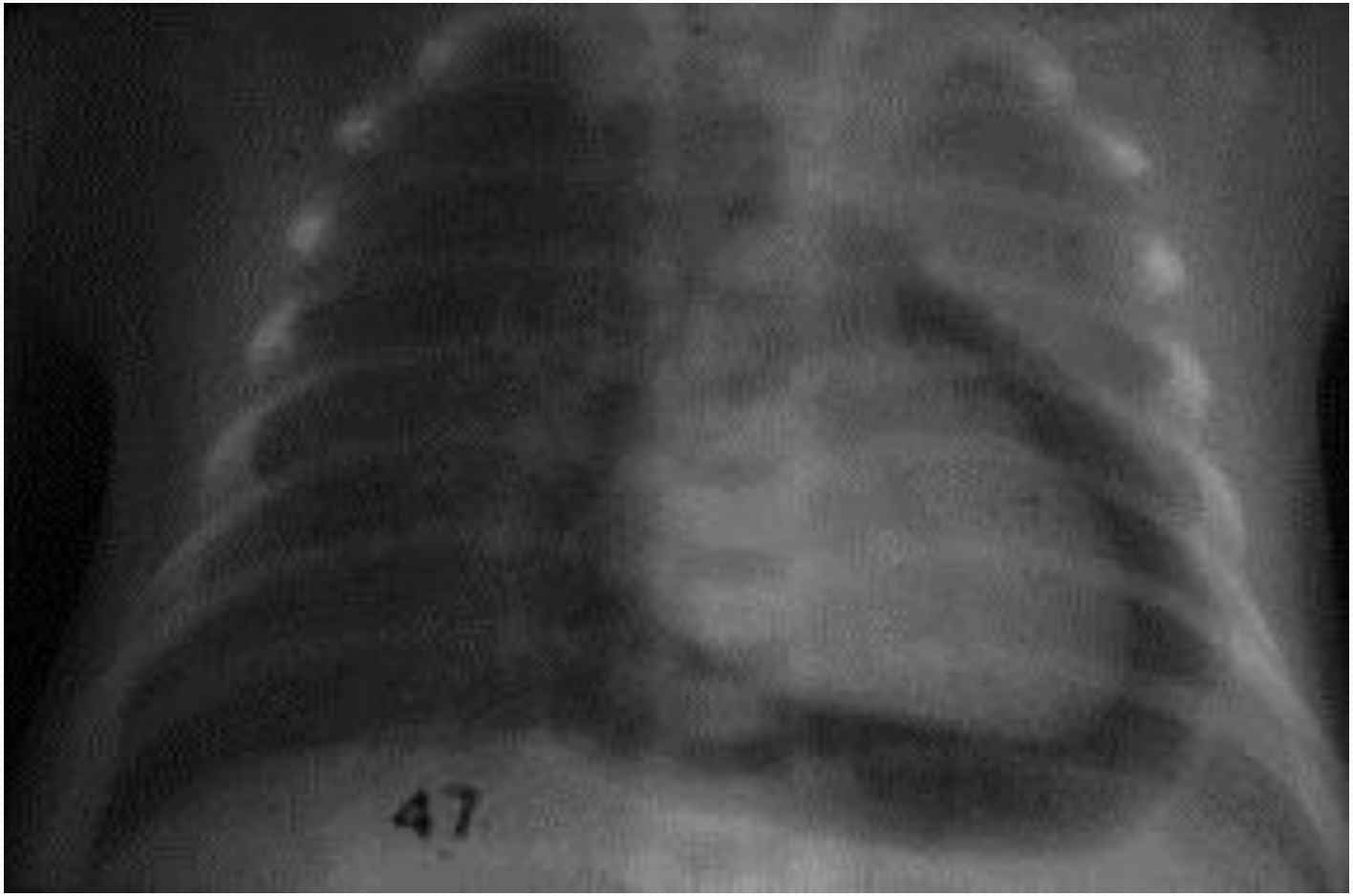
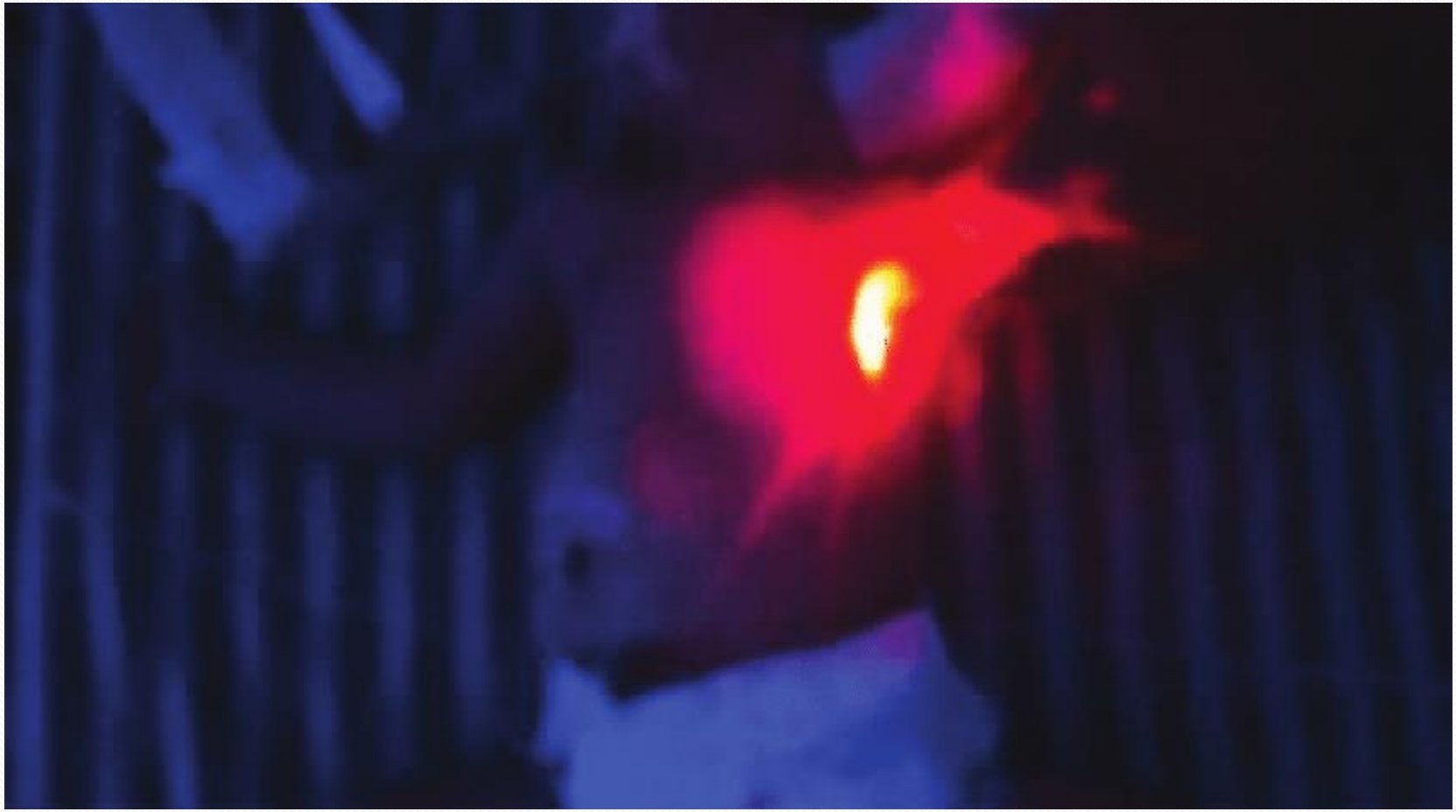


Image from Virtual Children's Hospital

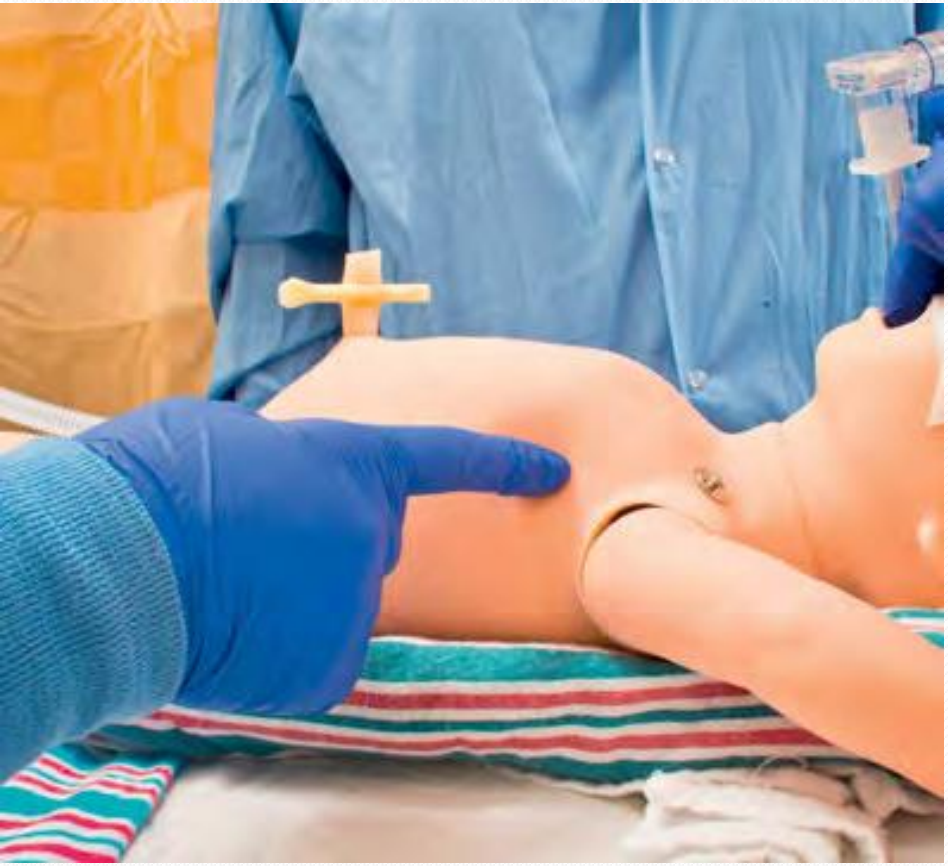
Diagnosis of pneumothorax

- transillumination of the chest
- decompression large pneumothorax (thoracocentesis)
- in stable neonates, radiographic confirmation should always be sought before intervention.

transillumination of the chest



Locations for percutaneous aspiration of a pneumothorax. Fourth intercostal space at the anterior axillary line (A), second intercostal space at the mid-clavicular line (B)



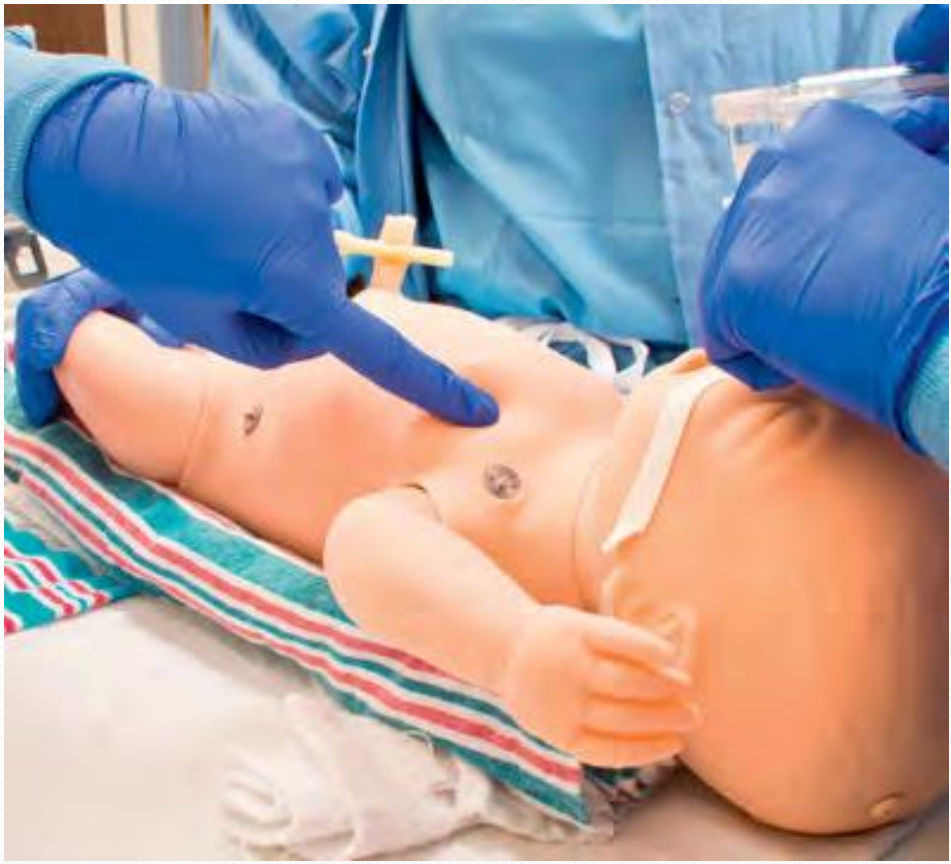
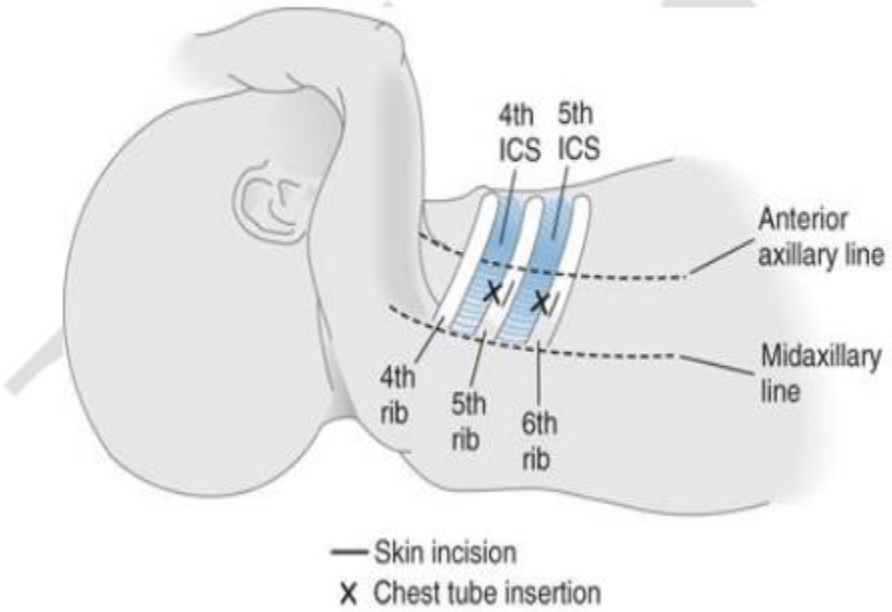
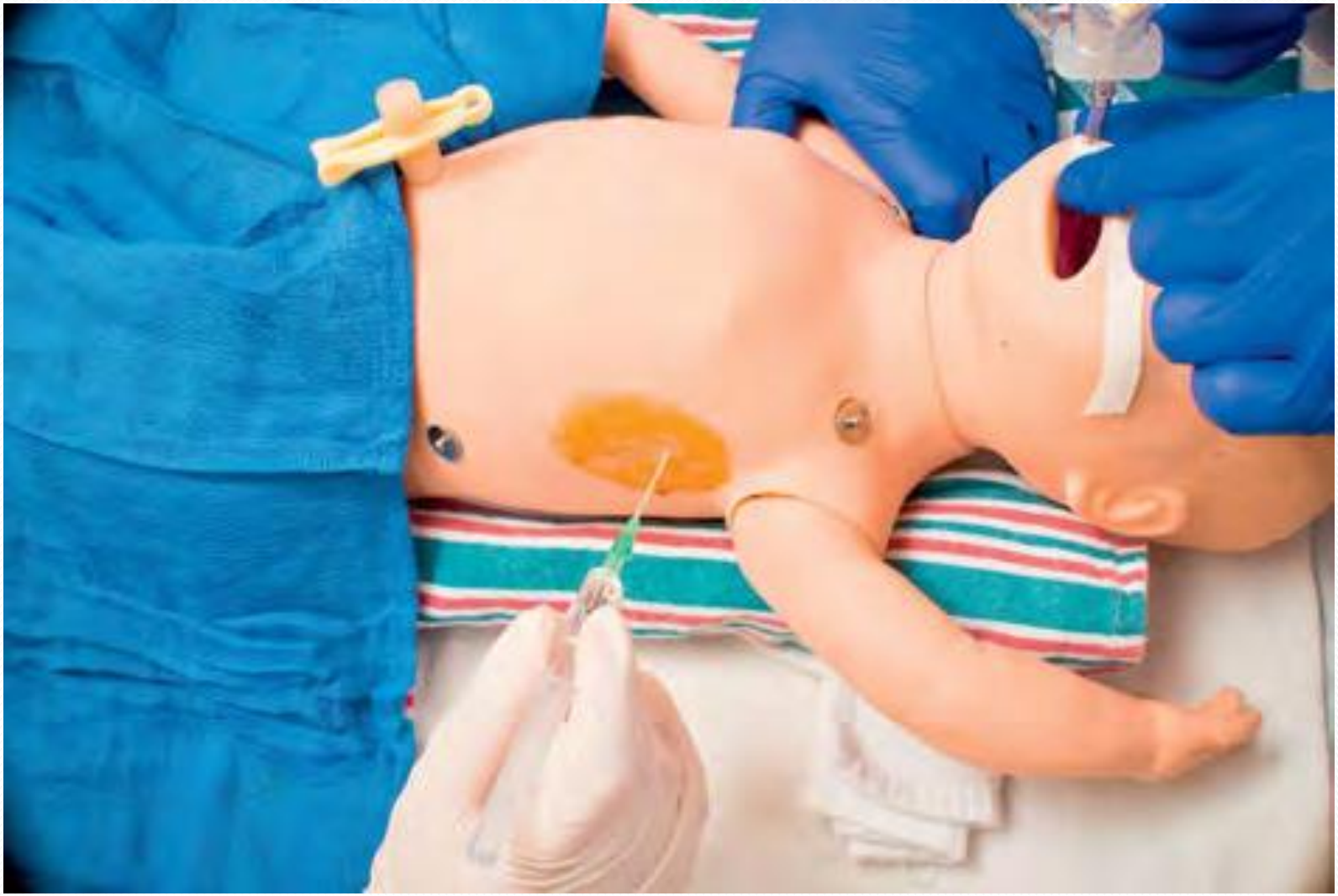


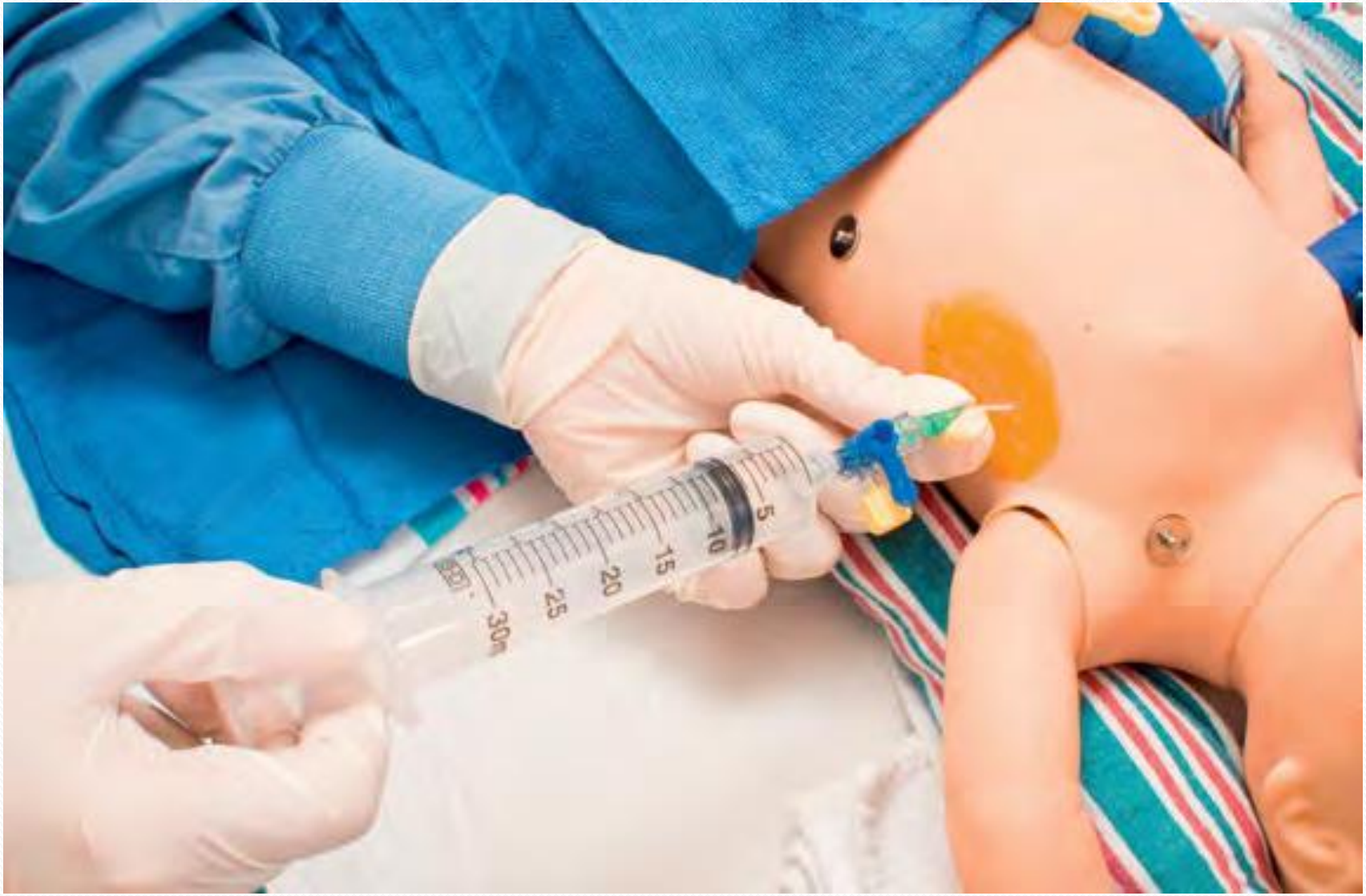
Image 1 : Anatomical reference points for insertion of a chest drain.



- Prepare the insertion site with topical antiseptic and sterile towels.
- Insert an 18- or 20-gauge percutaneous catheter-over-needle device perpendicular to the chest wall and just over the top of the rib.
- If an appropriate catheter-over-needle device is not available, a small “butterfly” needle may be used.
- The needle is placed over the top of the rib, rather than below the rib**, to avoid puncturing the blood vessels** located under each rib.

استفاده از شان پرفوره و رعایت استریلیته الزامی است





Management

- Treatment of a pneumothorax depends on the severity of clinical presentation and whether it occurs during mechanical ventilation.

- In asymptomatic patients without underlying pulmonary disease, no treatment is required; however, **close observation** for worsening pneumothorax and development of respiratory symptoms is clearly needed.
- The pneumothorax usually resolves spontaneously in 1-2 days.
- Follow-up radiographs can be obtained as mandated by the clinical picture.

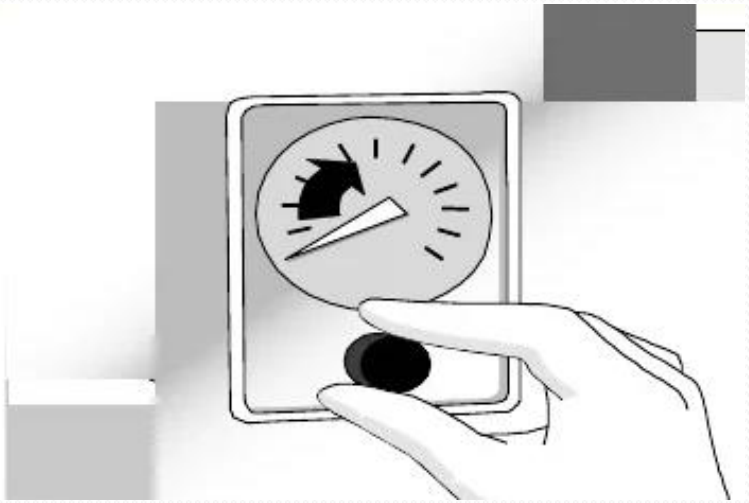
- In symptomatic patients with a mild pneumothorax without evidence of respiratory failure Only supportive care is needed.
- Theoretically, 100% oxygen given by a hood might lead to nitrogen washout and resolution or decrease in the size of the pneumothorax.
- Recent retrospective reviews showed **no difference** in time to resolution of pneumothorax in infants treated with 21% oxygen vs. 100% oxygen vs. somewhere in between
- **Oxygen is generally given only as needed to provide adequate oxygen saturations.**

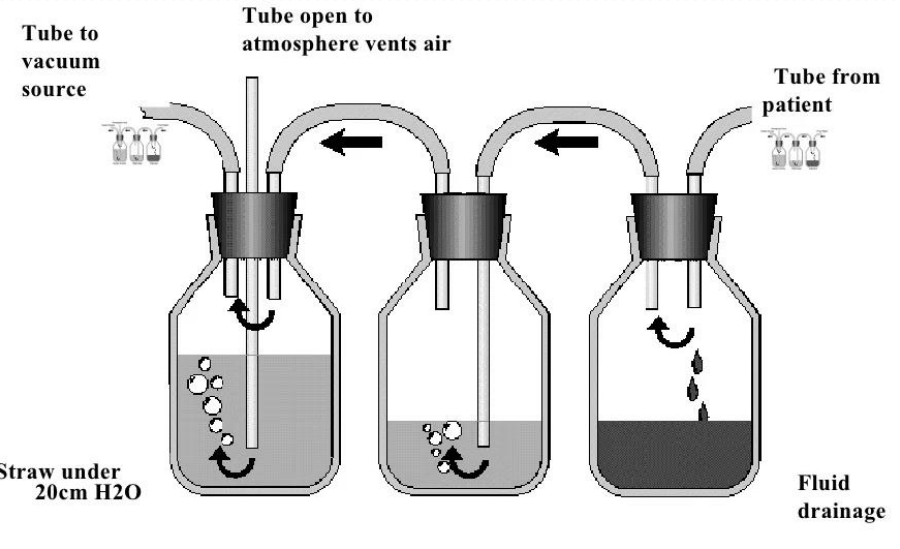
- **Thoracocentesis** is used for emergency evacuation of a large pneumothorax in unstable infants and might be the only treatment needed in nonventilated babies.

- However, recurrence of the pneumothorax should prompt the insertion of a chest tube.
- Thoracocentesis can also be used as a temporizing measure before chest tube insertion in ventilated infants.

- In ventilated patients with a large or tension pneumothorax, the initial management is to wean PIP and PEEP to decrease further air leak
- The position of the tube should be confirmed by anteroposterior and lateral chest radiographs.
- When placed correctly, there is immediate improvement in oxygenation in affected patients
- the tube should be connected to an underwater seal with **continuous suctioning at 10-20 cm H_2O**.

- The ***depth of the water*** in the suction bottle determines the amount of negative pressure that can be transmitted to the chest, ***NOT the reading on the vacuum regulator***



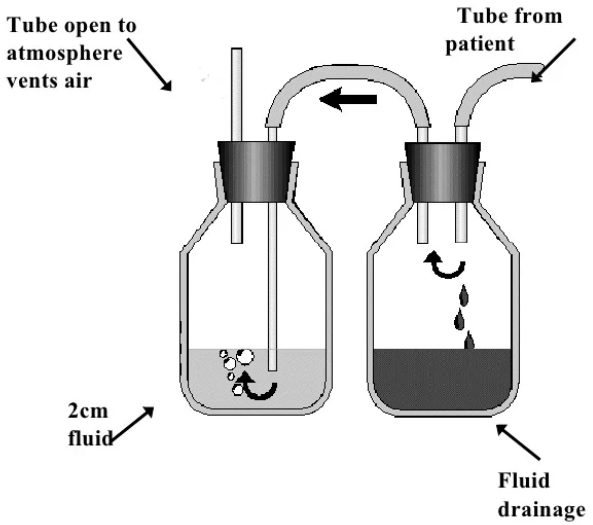


Collection bottle and water seal

Amount and rate of fluid drainage can be measured

Water seal remain fixed

Rely on gravity to create pressure gradient



- Once air bubbling stops, the chest tube should be clamped or put to water seal overnight, and if the pneumothorax does not reaccumulate, the tube can be successfully removed.