Chapter

Pneumothorax in Children

Hatice Sonay Yalçın Cömert

Abstract

Pneumothorax is a common pleural disease worldwide and is defined as the free accumulation of air between visceral and parietal pleura. Pneumothorax can be spontaneous, iatrogenic, and traumatic. Although it is less common than adults, it is seen in about 1.1–4 per 100,000 per year in the childhood age group. In patients presenting with variable clinic according to the cause of etiology, diagnosis is confirmed on a PA chest radiograph, sometimes a computed tomography may be required. The management of pneumothorax is varying from conservative, over intermediate (chest tube drainage) to invasive methods (video-assisted thoracoscopic surgery—VATS, thoracotomy). Here, we planned to write a chapter that includes a text containing general information about pediatric pneumothorax, algorithms, and visual and clinical cases of the causes of pneumothorax in children, including age, etiology, and treatment approach of pneumothorax in children.

Keywords: pneumothorax, children

1. Introduction

Although pneumothorax has been known in medical history since the times of Hippocrates and Galen, it was the first time that Itard named the term pneumothorax in 1803 [1]. Spontaneous pneumothorax due to bullae rupture was defined for the first time in 1926, and in 1932, Kjaergaard reported that pneumothorax may occur in completely healthy individuals due to isolated lung blebs [2]. In the treatment of pneumothorax, which was tried to be corrected with long bed rest, Noble has used a cannula, plastic drain, and underwater drainage system for the first time in 1873 [3]. The first thoracotomy and bulla resection was performed by Bigger in 1937, pleural abrasion by Churchill in 1941, subtotal parietal pleurectomy by Gaensler in 1956, and the first axillary thoracotomy and bulla excision and apical parietal pleurectomy by Deslauriers in 1980 [3].

2. Definition

Pneumothorax is defined as the free accumulation of air between visceral and parietal pleural space for various reasons. Pneumothorax can be spontaneous, iatrogenic, and traumatic in both neonatal and juvenile patients. Spontaneous pneumothorax is divided into two as primary and secondary. Primary spontaneous pneumothorax occurs secondary to apical blebs or bullae without evidence of other lung pathologies. Secondary spontaneous pneumothorax happens in the context of underlying lung diseases such as cystic fibrosis, asthma, connective tissue disorders, or pneumonia [4, 5].

Apart from these, if we define pneumothorax according to age, we should also mention neonatal and catamenial pneumothorax. Neonatal pneumothorax is the most common pneumothorax in childhood. It is reported that the cause is most likely the high transpulmonary pressure with the onset of breathing [6]. Catamenial pneumothorax is often associated with thoracic endometriosis syndrome.

3. Physiopathology of pneumothorax

Pressure in the pleural space is negative throughout the entire respiratory cycle, as the chest wall tends to expand and collapse in the lung. The pressure of −2 to −5 cm H_2O in expiration decreases to −25 to −30 cm H_2O in inspiration, and this pressure increases approximately 0.25 cm H_2O per cm from the lung basal to the apex [7]. Alveolar pressure is always greater than intrapleural pressure. Therefore, due to the high alveolar pressure and tension in the apical region, existing bleps and bullae in the apex may rupture. Thus, it causes air entry from the alveoli to the pleural space. Airflow continues until the pressure in the pleural space is equalized or until air leakage from the alveoli into the pleural space stops. This condition is called pneumothorax. Pneumothorax physiology includes a reduction in vital capacity and a decrease in oxygen partial pressure.

4. Types of pneumothoraces

Pneumothoraces can be classified as spontaneous (primary and secondary), iatrogenic, traumatic, neonatal, and catamenial pneumothorax. The types of pneumothoraces are shown in **Table 1**.

4.1 Spontaneous pneumothorax

Spontaneous pneumothorax (SP) is a comparatively rare condition in children. The peak age of occurrence in children is either in the neonatal period or in the late adolescent period [8]. Air enters the pleural space without any evident traumatic or iatrogenic mechanism. The incidence of pediatric SP is 4 per 100,000 in males and 1.1 per 100,000 in females with most occurring in patients 16–24 years of age [5, 9, 10]. SP is generally categorized into primary and secondary. In primary spontaneous pneumothorax (PSP), there is no underlying pathology and occurs unknown etiology. PSP refers to a pneumothorax from apical blebs or bullae [10]. However, secondary spontaneous pneumothoraces occur in children with underlying lung problems.

1. Spontaneous pneumothorax
a. Primary spontaneous pneumothorax
b. Secondary spontaneous pneumothorax
2. Iatrogenic pneumothorax
3. Traumatic pneumothorax
4. Neonatal pneumothorax
5. Catamenial pneumothorax

Table 1.
Types of pneumothoraces.

4.1.1 Primary spontaneous pneumothorax

A primary spontaneous pneumothorax (PSP) occurs without a precipitating event and in the absence of clinical lung disease and has an estimated incidence of 3.4 per 100,000 children with 4:1 male predilection [11]. In pediatric studies, the peak age of incidence occurs between 14 and 17 years of age, mainly in late teenagers [8]. The risk factors of PSP include tall and thin stature with low body weight [8]. Smoking is also the primary environmental risk factor for primary spontaneous pneumothorax, especially in teenage patients [12]. Some studies have shown that familial and genetic forms of PSP are related to mutations in the folliculin gene on chromosome 17 in the literatüre [5, 12].

It has been recommended that subpleural blebs and bullae are causally related to the development of primary SP and may be clarified by that these tall and slim children tend to have higher transpulmonary pressure at lung apex, and their rapid growth relative to pulmonary vasculature may result in ischemia and thus blebs evolution at these regions [5, 8].

Most patients are clinically stable on initial evaluation and small cases may present in fulminant distress [1]. Chest pain and shortness of breath are common presenting symptoms of PSP and may be developed at rest or accelerated by any maneuver that increases intrathoracic pressure (Valsalva) [5, 13]. Other clinical findings in patients with pneumothorax include cough, ipsilateral hypoventilation, and nonspecific respiratory distress [4, 5].

Sample chest X-ray and thorax-computed tomography of our patients admitted with primary spontaneous pneumothorax from our archive are shown in **Figures 1** and **2**.

4.1.2 Secondary spontaneous pneumothorax

Commonly known situations predisposing individuals to a secondary spontaneous pneumothorax (SSP) include primary lung disease as asthma, cystic fibrosis, interstitial emphysema, inflammatory/connective tissue diseases such as Marfan syndrome, Ehlers-Danlos syndrome, juvenile idiopathic arthritis, systemic lupus erythematosus, polymyositis, dermatomyositis, sarcoidosis, Langerhans cell histiocytosis, α_1-antitrypsin deficiency, Birt–Hogg–Dube syndrome, infections such

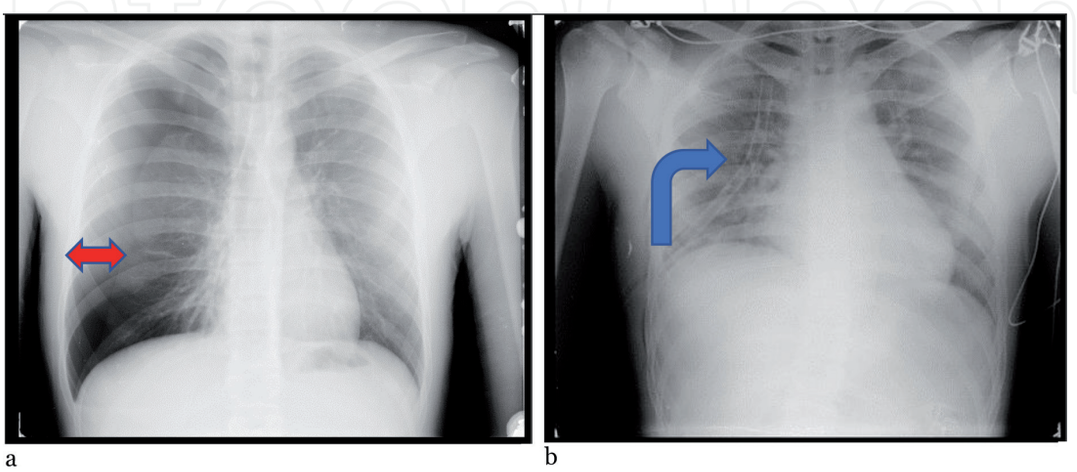

Figure 1.
(a) A 16-year-old male patient presented to the emergency department with a sudden onset of chest pain and was diagnosed with spontaneous pneumothorax on the right side of his chest X-ray (free air in the thorax marked with a red arrow). (b) Film of the same patient after right side chest tube placement (inserted chest tube marked with blue arrow).

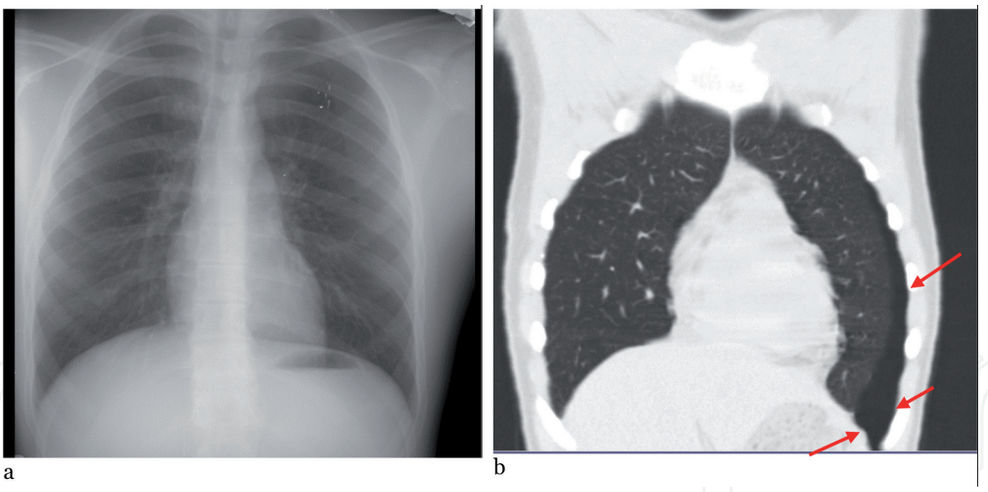

Figure 2.
A 17-year-old male patient presented to the emergency department with the complaint of sudden onset of chest pain. (a) There was no pneumothorax in the anterior–posterior chest X-ray of the patient. (b) Minimal pneumothorax image on the left side in the thorax-computed tomography of the patient (free air in the thorax marked with red arrows).

Primary lung disease: asthma, cystic fibrosis, interstitial emphysema	Infection: *Pneumocystis jirovecii*, tuberculosis, necrotizing pneumonia/abscess, measles, human immunodeficiency virus/acquired immunodeficiency syndrome, parasitic
Inflammatory/connective tissue disease: Marfan syndrome, Ehlers-Danlos syndrome, juvenile idiopathic arthritis, systemic lupus erythematosus, polymyositis, dermatomyositis, sarcoidosis, Langerhans cell histiocytosis, α_1-antitrypsin deficiency, Birt–Hogg–Dube syndrome	Malignancy—lymphoma, metastases
Foreign body aspiration	Congenital malformation: congenital cystic adenomatoid malformation, congenital lobar emphysema

Adapted from Dotson et al. [5].

Table 2.
Causes of pediatric secondary spontaneous pneumothorax.

as *Pneumocystis jirovecii*, tuberculosis, necrotizing pneumonia/abscess, measles, human immunodeficiency virus/acquired immunodeficiency syndrome, parasitic, malignancy (lymphoma, metastases), foreign body aspiration, and congenital malformations such as congenital cystic adenomatoid malformation and congenital lobar emphysema [5, 14]. SSP causes are summarized in **Table 2** [5].

The theorized mechanism is chronic airway inflammation that causes small airway obstructions and creates the pressure needed for air to escape into the pleural space. These conditions can make the lung pleura more susceptible to rupture and subsequent development of pneumothorax [15]. The most important symptom of SSP is dyspnea, tachypnea, and tachycardia.

Cystic fibrosis (CF) is a severe obstructive airways disease and one of the most common causes of secondary spontaneous pneumothorax. Pneumothorax is seen approximately 3.4% of all patients will suffer from CF during their lifetime and mostly occurs in adult patients [16, 17]. Cysts, blebs, and bullae are all commonly found in the lungs of CF patients, and these cause gas to accumulate in the small airways, resulting in a cystic appearance. The typical presentation is acute onset of chest pain and

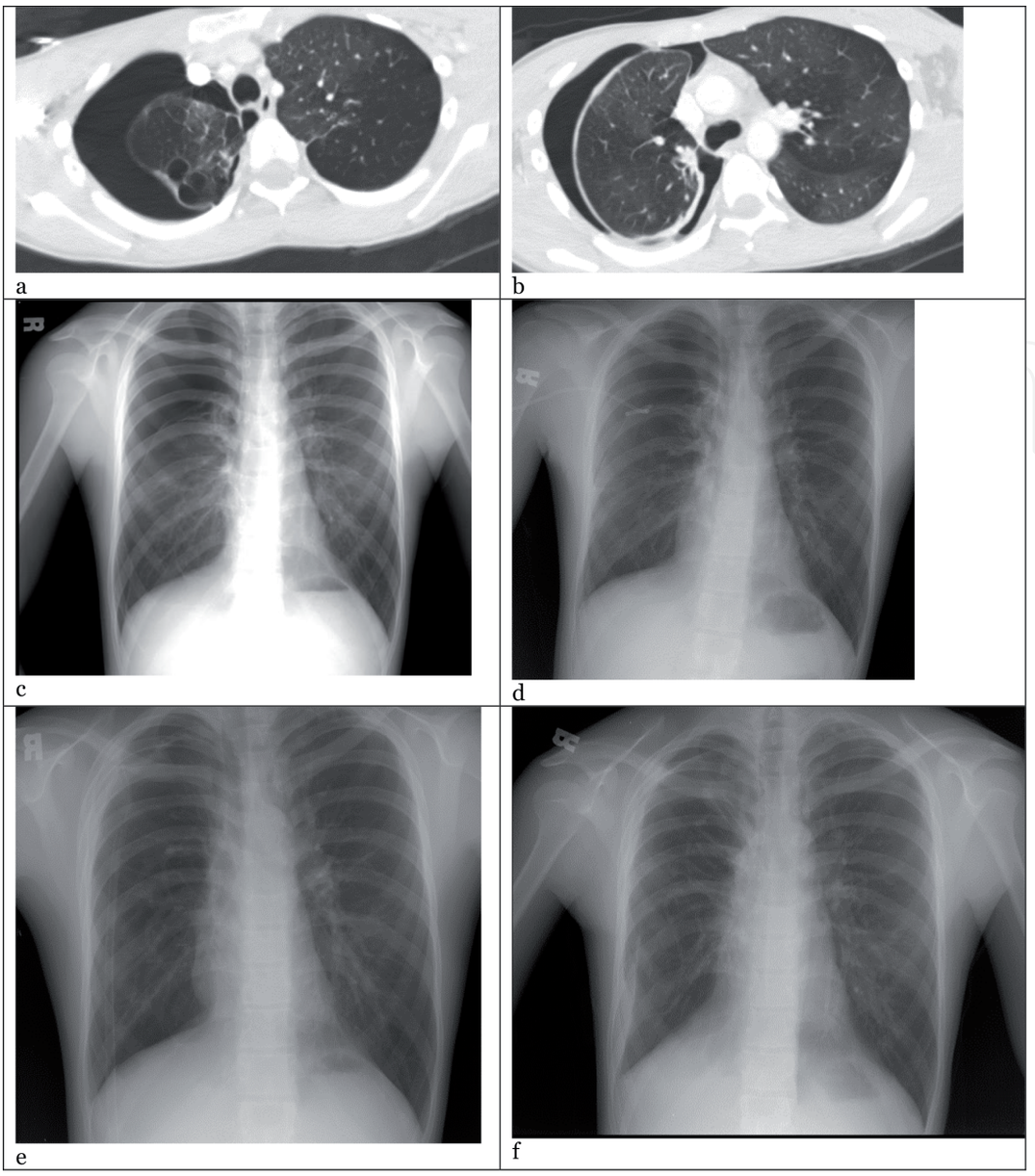

Figure 3.
A 13-year-old male patient followed up with the diagnosis of cystic fibrosis was admitted to the emergency department with respiratory distress. Upon the presence of pneumothorax in the right upper lobe in the thorax in computed tomography (a, b) and the chest X-ray (c), a pig-tail catheter was placed in the right thorax (d). The child's clinical condition did not improve and thoracotomy with pleurectomy was performed. The child was followed up with a chest tube after the operation (e), and has been covered and discharged (f).

breathlessness, and the treatment decisions include the size of the pneumothorax, severity of disease, stability of the patient, and whether this is the first or a recurrent pneumothorax [16]. Pneumothorax due to cystic fibrosis can also be seen in the childhood age group and invasive surgeries may be required. A spontaneous pneumothorax chest X-ray film of a CF patient from our archive is shown in **Figure 3a–f**.

4.2 Iatrogenic pneumothorax

The most frequent cause of iatrogenic pneumothorax is a transthoracic pulmonary biopsy, but it also may appear as a complication of many other procedures and caused by barotrauma secondary to mechanical ventilation [18, 19]. Iatrogenic pneumothorax is related to underlying lung disease along with high ventilatory settings [19]. The most common cause of iatrogenic pneumothorax is invasive diagnostic and therapeutic procedures, such as central venous access, thoracocentesis,

thoracic surgery, or intubation [1]. Iatrogenic pneumothorax may also develop during cardiopulmonary resuscitation and tracheostomy.

4.3 Traumatic pneumothorax

Although thoracic injuries occur less frequently in children than adults, thoracic trauma in children carries a 5% mortality [20, 21]. The most causes of trauma in pediatric patients are traffic accidents, followed by falling from heights, and bicycle accidents [22]. The greater flexibility of the thoracic cage in young children permits the anterior ribs to be compressed to meet the posterior ribs [23]. Because of the flexibility, pulmonary contusions are more common than rib fractures in children [23].

The most common injury in children with blunt thoracic trauma is pulmonary contusion and pneumothorax, which is observed as isolated injury in 30% of the cases [22].

Traumatic pneumothorax can be classified as small occult, tension, and open (**Table 3**). A small pneumothorax from blunt torso trauma is often asymptomatic, with more than half identified as being occult (defined as a pneumothorax observed on computed tomography scan of the chest, but not on chest radiograph) [22]. However, a large pneumothorax may cause clinical symptoms that overlap with those produced by lung parenchymal damage—tachypnea, distress, and decreased saturation [22]. A traumatic pneumothorax and contusion chest X-ray film of a patient from our archive is shown in **Figure 4**.

Open pneumothorax	Related to an open chest wall injury
Occult pneumothorax	Small pneumothorax without clinical significance, typically seen in trauma
Tension pneumothorax	Rapid accumulation of air within the thoracic cavity that leads to a reduction in central venous return as well as tamponade effect on cardiac output

Table 3.
Characteristics of traumatic pneumothorax [14].

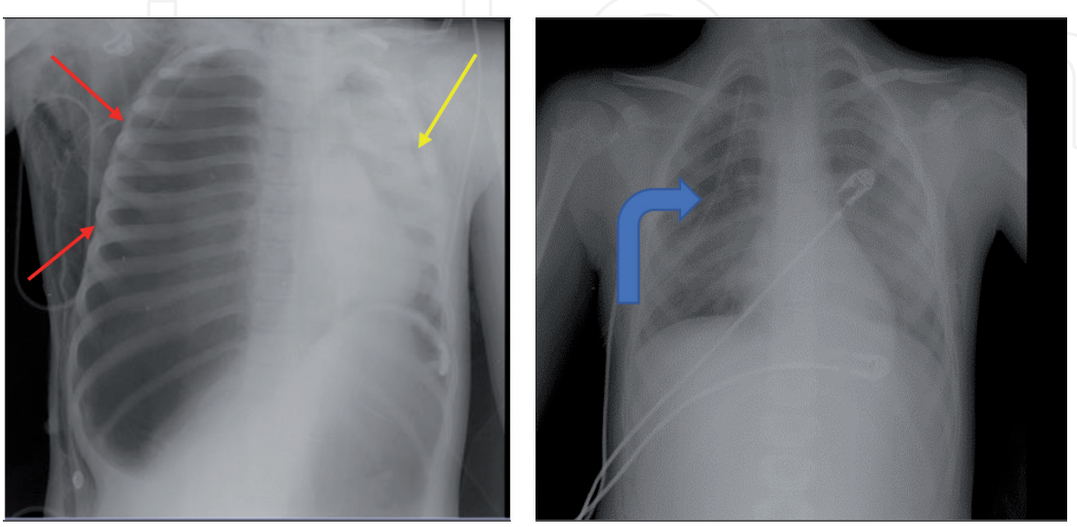

Figure 4.
An 8-year-old male patient applied to the emergency department due to a traffic accident. (a) Pneumothorax in the right thorax and contusion in the left lung were detected in the chest X-ray (free air in the right thorax marked with red arrows and contusion has shown with yellow arrow). (b) Chest X-ray after tube placement in the patient's right thorax (inserted chest tube marked with blue arrow).

When the mediastinum is displaced to the contralateral side with impairment of the venous return, the tension pneumothorax occurs and is more common in children [22]. The symptoms of tension pneumothorax are tachycardia, severe respiratory distress, and hypoxemia, with hypotension and tracheal deviation. Heartbeat is heard on the opposite side and the neck veins become dilated and severe cyanosis occurs. No chest X-ray is required to insert a chest tube in children with tension pneumothorax. The child's symptoms improve dramatically with chest tube insertion.

Open pneumothorax is usually seen after penetrating injuries. This causes a collapse in the lung on the side of the trauma and ventilation failure in the other lung. The patient who develops open pneumothorax is cyanosed and has serious respiratory distress is present. In the treatment, the defect should be closed with a sterile gas.

4.4 Neonatal pneumothorax

Neonatal pneumothorax, with an incidence of 1–2% in newborns, is symptomatic in 0.08% of all live births and is reported as 5–7% in those with a birth weight of less than 1500 g, although it can reach 30% in those with an underlying lung problem and those who need mechanical ventilation comes out [10, 11]. The most common cause of this condition is barotrauma [13]. In addition, male gender and cesarean delivery are also considered among risk factors [11].

In order to inflate the lungs of a newborn baby when he is not breathing himself, mechanical ventilation with an average pressure of 50–80 cm H_2O is required to overcome the high transpleural pressure. During this resuscitation, the air given into the lungs is distributed with an uneven pressure inside the lungs. As a result, some alveoli are ruptured and air passes from the peribronchial area to the mediastinum and pneumothorax develops [24]. A chest X-ray visualization from a newborn from our archive who needed resuscitation at 41-week postpartum and had pneumothorax on the right in the chest X-ray has been shown in **Figure 5**.

It most commonly occurs in the first three days and should be suspected in cases of sudden respiratory distress, decrease in oxygen saturation, inability to listen to breath sounds, or when ventilator parameters have to be increased.

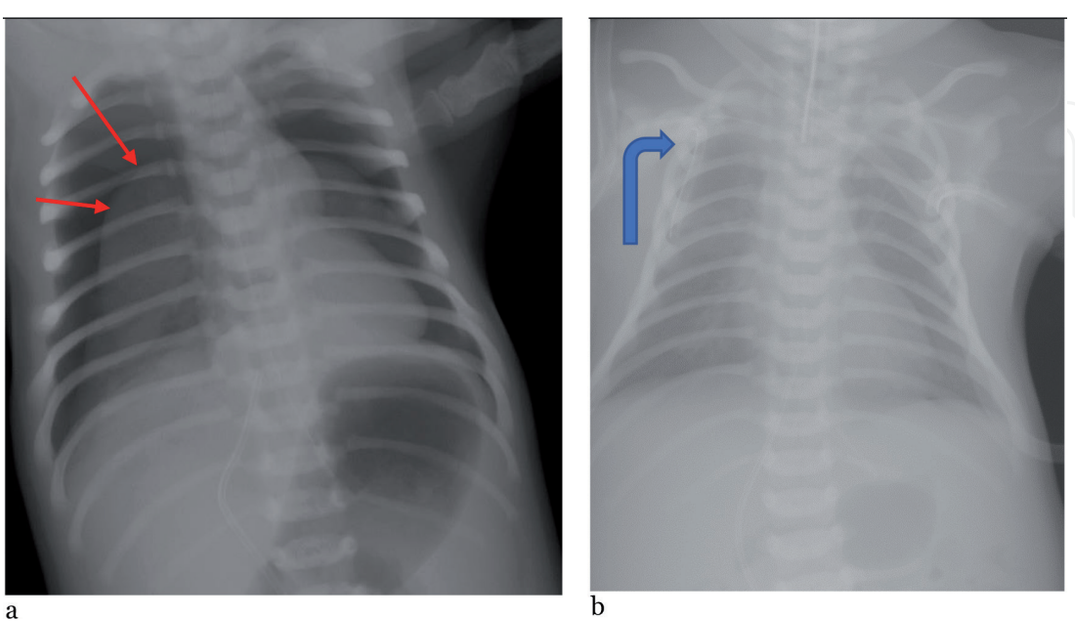

Figure 5.
(a) Film of newborn who needed resuscitation at 41-week postpartum and had pneumothorax on the right in the chest X-ray (free air in the thorax marked with red arrows). (b) Film of the same newborn after right-side chest tube placement (inserted chest tube marked with blue arrow).

It causes high mortality and morbidity, especially in premature babies and newborns with underlying lung parenchyma disease. Whatever the cause, neonatal pneumothorax needs to be treated very quickly because pneumothorax in neonates will lead to serious complications, including lung perforation, phrenic nerve palsy, chylothorax, and hemopericardium [11].

4.5 Catamenial pneumothorax

Catamenial pneumothorax (CP) is a form of thoracic endometriosis syndrome, which also includes catamenial hemothorax, catamenial hemoptysis, catamenial hemopneumothorax, and endometriosis lung nodules, as well as some exceptional presentations [25]. The most common extrapelvic manifestation of endometriosis is thoracic endometriosis and often presents as catamenial pneumothorax [10]. Most commonly occurs in women aged 30–40 years, but has been diagnosed in young girls as early as 10 years of age and postmenopausal women (exclusively in women of menstrual age) most with a history of pelvic endometriosis [25].

CP is a rare and important condition of recurrent pneumothoraces, which occurs within 48–72 h from the onset of menses [11]. The pathophysiology is not completely understood but it is treated with hormonal therapies [11].

5. Diagnosis

The diagnosis of pneumothorax can be made by physical examination or imaging studies including chest X-ray, ultrasonography, and computed tomography (CT) scan [19]. A conventional chest X-ray is a typical imaging examination used to confirm the diagnosis of pneumothorax and a CT scan may be validated to show smaller pneumothoraces. CT scan is commonly accepted as the gold standard in pneumothorax diagnosis [1]. Dotson et al. proposed that detection of blebs/bullae on the CT scan may be predictive of recurrence of PSP, especially bilaterally pneumothoraces [5]. There are multiple methods for calculating pneumothorax sizes like Light, Rhea, and Collins for adults, but these methods are not appropriate for the childhood age group [5, 26, 27].

Dahmarde et al. suggested that ultrasound is accurate and reliable for newborn pneumothoraces [28]. Ultrasound can result in timely diagnoses specifically in neonatal pneumothorax and facilitates the therapy process; lack of ionizing radiation and easy operation are the benefits of this imaging technique.

6. Treatment

The treatment options are changing by age, size, and the type of pneumothorax in childhood. There are no standardized guidelines for therapeutic interventions for children with pneumothorax; however, early identification and appropriate management can reduce morbidity and mortality.

While the size of the pneumothorax can be calculated by various methods in adults, there is no method that can be applied to children yet. Although minimal pneumothoraxes that do not cause clinical problems can be followed conservatively, most patients require drainage, and a thorax tube is inserted. Nonoperative treatment methods are monitoring with supplemental oxygen (100% high-flow) or needle aspiration. Surgical treatment methods range from the insertion of a chest tube to more invasive interventions such as video-assisted thoracoscopic surgery (VATS) or thoracotomy, including resections, pleurodesis, or bullectomy [10]. Surgical

Children weight (kg)	Tube size (French)
<3	8–10
3–8	10–12
8–15	12–16
16–40	16–20
>40	20–24

Table 4.
Guide for chest tube selection for pneumothorax [4].

indications for pneumothorax are resistant and prolonged air leak (>4 days), persistent and recurrent pneumothorax, large pneumothorax, first pneumothorax with a history of pneumothorax in the other lung, and bilateral pneumothorax [10, 11]. Chest tube placement should be the first choice in patients with surgical indication, and then, open or closed surgical techniques should be planned according to the child's clinic. Although the VATS procedure is easily used in the childhood age group, thoracotomy with resections, pleurodesis, or bullectomy may be preferred or needed in cases with severe air leak and recurrent pneumothorax [29].

Pleural catheters are tools that are placed in the fourth, fifth, or sixth intercostal space in generally anterior or midaxillary line with Seldinger technique and placed to water seal in children. In newborns, the catheters are usually placed from the second or third midclavicular line with again Seldinger technique and placed to water seal. The chest tube sizes are changing from the patient's size and age. The guide for chest tube selection for pneumothorax for children patients is summarized in **Table 4** [4].

The aim of surgical treatment is to resect of blebs and bullae and pleurodesis to prevent recurrences. VATS procedure is performed with good results in children with PSP and as the gold standard for surgical management of PSP by using various surgical instruments from 1 to 3 incisions of approximately 1.5–2 cm, which are opened on the chest with the help of a video [29, 30]. Blebs and bullae due to pneumothorax are removed with VATS with the help of staples. Pleurodesis ensures that the parietal and visceral pleura sheets stick together. Pleurodesis can be performed by using pleurectomy, pleural abrasion, or chemicals [31].

Lewit et al. suggested that nonoperative methods are not suitable for the treatment of pneumothorax and mentioned a decreased recurrence rate in those undergoing surgical treatment at initial presentation in the childhood age group [32]. Also, Lopez et al. have observed decreased median total length of stay and decreased recurrence rate in the surgical group compared with the initial non-VATS group in children [11].

On the other hand, Brown et al. discussed whether conservative management is an acceptable alternative to nonconservative procedures and found that conservative management of primary spontaneous pneumothorax was similar to interventional management, with a lower risk of significant adverse events [33].

The general approach is chest X-ray negative and CT scan positive pneumothoraces do not require invasive methods and they can be followed conservatively [23]. However, since childhood is a wide range, a pneumothorax that looks small may even be mortal for a newborn premature baby. Although thoracic tube insertion is a minor surgical procedure, every procedure has surgical stress, especially for neonatal intensive care patients. Therefore, every child with pneumothorax for whom a follow-up decision is made requires very special close follow-up. Likewise, close follow-up of a child patient with a chest tube placed should be very important in terms of possible complications.

7. Complications

The most common complications of pneumothorax seen in childhood are air leak, tension pneumothorax, pneumomediastinum, subcutaneous emphysema, hemothorax, and very rarely Horner's syndrome. If the air leak is continued within 48 hours of pneumothorax treatment, it may become resistant. Therefore, a second chest tube or even VATS or thoracotomy may be required for persistent air leakage, depending on the age of the child or the etiology of the pneumothorax [3].

8. Conclusion

In conclusion, the etiology and management vary according to age and type of pneumothorax in the childhood age group, and this is a life-threatening special condition that requires urgent intervention and special follow-up.

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