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ORIGINAL ARTICLE

Etiologies of Spontaneous Pneumomediastinum in Children of Different Ages

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Background: Spontaneous pneumomediastinum (SPM), while rare, is probably underestimated in children. Treatments for SPM target the underlying disease and trigger factors. This study aimed to analyze the etiology of SPM in different age groups.

Methods: A total of 18 children with SPM were analyzed in the Children's Medical Center at China Medical University Hospital between 1997 and 2007.

Results: The incidence of SPM in children was 1:8,302 patients at the Department of Pediatric Emergency Medicine. A bimodal peak in incidence occurred in those younger than 4 years old and in those aged 15–18 years. Characteristic symptoms were dyspnea (77.8%), followed by chest pain (66.7%) and neck pain (44.4%); common specific physical signs were subcutaneous emphysema (55.6%) and Hammer's sign (11.1%). Trigger factors were infection (44.4%), with a mean age of 5.8 ± 5.0 years, and diabetic ketoacidosis (5.6%), with a mean age of 18 years. Idiopathic SPM accounted for 50.0% of patients, with mean age 14.4 ± 1.8 years. In terms of the age distribution, five (27.8%; males/females=4:1) preschoolers (<6 years old) developed SPM, mostly due to infectious disease. Two girls aged less than 10 years developed asthma in later years. All eight male adolescents (≥ 10 years) developed SPM due to idiopathic factors. Of nine boys with idiopathic SPM, six underwent strenuous exercise before developing SPM. Mean hospitalization was 7.9 ± 11.5 days and 11 (61.1%) patients needed intensive care. Nearly all of the patients had complete resolution on chest radiography before discharge.

Conclusion: Clinicians should be alert to the risk of SPM based on the presence of these symptoms. The etiology of SPM varies with age. Treatment of SPM must target the trigger factors or the underlying disease.

1. Introduction

AQ1 Spontaneous pneumomediastinum (SPM) is a syndrome that refers to patients with mediastinal air leak without chest trauma, mechanical ventilation, iatrogenic procedure or surgery. SPM can occur due to air leakage from an intra- or extrathoracic source,^{1,2} but rarely before the age of 18 years.^{1,3} In

most cases, air leakage from ruptured alveoli escapes and dissects the hilum along the peribronchovascular sheaths and spreads to the mediastinum.^{4,5} Once in the mediastinum, air extends around the large vessels and the esophagus to the thoracic wall. Chest radiographic signs that indicate pneumomediastinum (PM) include a vertical radiopaque streak along the left side of the heart

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and aortic arch, continuous diaphragm signs, and radiopaque streaks along retrosternal, pericardiac and peritracheal areas, which are logical according to the pathogenesis of PM. However, SPM was diagnosed by chance on chest radiography in one of our patients with isolated chest pain. Children with SPM might present with chest pain, subcutaneous emphysema, dyspnea and Hamman's sign. Rarely, tension PM leads to airway compression and venous obstruction.

SPM was first described by Hamman in 1939,⁶ and has become increasingly recognized as a distinct clinical entity. Most studies on PM have been conducted in adults; few studies have been conducted in children. Possible precipitating or trigger factors were reported in 70–90% of cases from previous studies.^{3,9,10} Respiratory and Valsalva maneuvers, such as coughing, crying-screaming, vomiting, hypercapnea, competitive athletics and inhalation of illicit drugs, were cited as causes in children. Medical conditions such as asthma, bronchiolitis, bronchopneumonia, convulsion or laryngitis are also associated with PM.¹⁵ This study aimed to investigate the characteristics of clinical presentation, precipitating factors, hospital course and possible sequelae in children of different ages.

2. Materials and Methods

2.1. Patients

This was a retrospective chart review of 26 patients with PM aged 6 months to 18 years of age who attended China Medical University Hospital (CMUH) between November 1997 and July 2007 according to the international classification of discharge code. The review did not cover neonatal PM, which might be considered as a separate entity. Eight patients with PM occurring in the setting of ventilator use, trauma, iatrogenic procedure, surgery or with history of underlying lung disease were excluded from further analysis. Therefore, a total of 18 patients were monitored at CMUH and included in this analysis.

2.2. Methods

AQ2 Diagnosis was made using **posteroanterior** (PA) or AQ3 **anteroposterior** (AP) chest radiographs, including the cervical region. Adding lateral chest films considerably improved the diagnostic accuracy. For each patient with SPM, basic data, past history, presenting symptoms and signs, possible precipitating factors, associated complications, hospital course and outcome were collected. Possible causes and precipitating factors were collected according to

presenting symptoms and signs. The review of medical records and history focused on identifying predisposition or trigger factors such as infectious disease, medical condition or strenuous sports, which often involve a vigorous Valsalva maneuver. The presenting symptoms and signs were also collected, including dyspnea, chest pain, neck pain, sore throat, nausea, vomiting, dysphagia, back pain, shoulder pain, subcutaneous emphysema (SCE) or Hammer's sign. Hammer's sign is defined as a crunching noise, made by air in mediastinal tissues, and is almost pathognomonic for this condition and, at times, linked with a reduction in heart sounds. Previously healthy children who developed SPM without apparent predisposing or trigger factors were classified in the idiopathic group. For suspected esophageal rupture, esophagography was performed. Severity could be related to the volume of escaping air into the mediastinum, airway compression with respiratory distress and obstruction of venous return with pseudotamponade. Intensive care should be considered under impending decompensation. The length of stay and the incidence of intensive care unit (ICU) care were also analyzed.

2.3. Statistical analysis

Descriptive data are reported as means±standard deviation and analyzed by SAS 9.1 (SAS Institute, Cary, NC, USA) statistical software. Wilcoxon rank-sum test and Fisher's exact test were used for categorical variables. A *p*-value < 0.05 was considered statistically significant.

3. Results

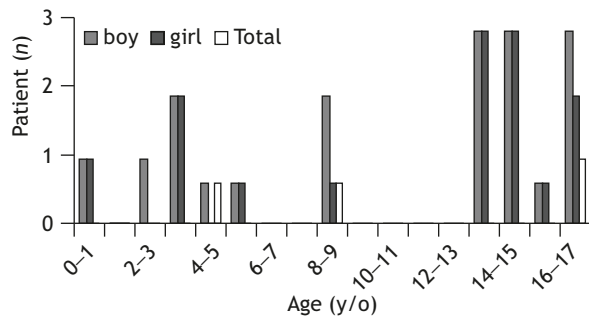
3.1. Characteristics of patients with SPM

The mean age of the 14 boys (77.8%) and four girls (22.2%) with SPM was 10.8±5.8 years, with a range of 9 months to 18 years. None smoked or used drugs. The presence of SPM was confirmed radiographically in all patients. Bimodal peaks in incidence occurred in children aged less than 4 years and in those aged 15–18 years (Figure 1). Boys were predominantly affected (*p*=0.0294). The age distribution of SPM in boys was compatible with the bimodal peak. The limited number of girls posed difficulty in categorizing the age distribution.

3.2. Etiology

Table 1 summarizes the clinical course and demographics. The most common causes in these patients was infection (8/18, 44.4%). Nine patients (9/18, 50.0%) were identified as the idiopathic group.

One patient had SPM complicated with diabetic ketoacidosis (DKA) (1/18, 5.6%). None of the cases were due to foreign body aspiration or esophageal rupture. In the infectious disease group, bronchopneumonia and bronchiolitis were the two leading causes. Most patients had wheezing in breath sounds on physical examination; two (11.1%) girls developed asthma later, manifesting SPM in the first asthma attack. In terms of age distribution, infection was the leading cause of SPM in preschool age and middle childhood (mean: 5.8 ± 5.0 years). In five preschoolers (<6 years), four boys and one girl had infectious disease. In adolescents (≥ 10 years), idiopathic SPM was most common (mean: 12.1 ± 4.3 years, $p=0.0022$). Six (6/10, 60.0%) of the male adolescents developed SPM after strenuous exercise.



AQ4 **Figure 1** Age distribution of spontaneous pneumomediastinum in children

3.3. Clinical findings

Table 2 shows the associated presenting symptoms and signs, predominantly dyspnea and chest pain. The latter worsened with deep respiration and postural change in seven patients (7/18, 38.9%) and radiated to the back, shoulder or arms in four patients (22.2%). The most common clinical sign was SCE (55.6%). Two (11.1%) patients had Hamman's sign and one (5.6%) patient had both SCE and Hamman's sign. Six patients (33.3%) had neither SCE nor Hamman's sign.

3.4. Hospital courses

Table 3 shows the hospital course. All patients were admitted to the hospital; 11 patients (61.1%) required intensive care due to respiratory distress. Seven patients (7/11, 63.6%), aged 4.4 ± 2.8 years, had bronchopneumonia and four patients (4/11, 36.4%), aged 14 ± 2.7 years, had idiopathic SPM. The length of stay in the ICU was 10.4 ± 11.8 days and 1.5 ± 0.87 days, respectively ($p=0.0197$). Risk factors for intensive care were young age and bronchopneumonia. The mean duration of hospitalization of all cases was 7.9 ± 11.5 days, range 2–53 days. The idiopathic group were hospitalized for 4.0 ± 2.6 days and the infection group for 13.0 ± 16.0 days ($p=0.0156$). A 9-month-old male infant had the longest hospital stay (53 days) due

Table 1 Clinical causes and demographics of spontaneous pneumomediastinum in children

	No. (%)	Age (yr), mean	SD	Sex (M:F)
Infections	8 (44.4)	5.8	4.9	4:4
Bronchopneumonia	7	6.4	5.1	4:3
Bronchiolitis	1	2	0	1:0
DKA	1 (5.6)	18	0	1:0
Idiopathic	9 (50.0)	12.1	4.3	9:0
Total	18 (100.0)	10.8	5.8	14:4

DKA = diabetic ketoacidosis; F = females; M = males; SD = standard deviation.

Table 2 Clinical presentation of spontaneous pneumomediastinum in 18 children

Symptoms	n (%)	Signs	n (%)
Dyspnea	14 (77.8)	SCE	10 (55.6)
Chest pain	12 (67.7)	Hamman's sign	1 (11.1)
Neck pain	8 (44.4)	Neither	6 (33.3)
Sore throat	6 (33.3)	Both	1 (5.6)
Back pain	3 (16.7)		
Dysphagia	1 (5.6)		
Shoulder pain	1 (5.6)		

SCE = subcutaneous emphysema.

to bronchopneumonia concomitant with pneumothorax, PM and respiratory failure.

3.5. Treatment and outcome

The primary treatments were primary analgesics and management of the underlying condition. Chest pain was relieved by acetaminophen and/or nonsteroidal anti-inflammatory drugs. Bronchodilators or prokinetic agents were prescribed for apparent wheezing, asthma attack and/or vigorous vomiting. Rest and oxygen therapy were prescribed for all patients to prevent the progression to pneumothorax and respiratory distress.

3.6. Complications

Three boys (3/18, 16.7%) aged 6.9 ± 6.0 years had concomitant PM, pneumothorax and dyspnea; all received chest tube insertion, while two underwent endotracheal tube insertion and ventilation. Two patients (25.0%) in the infection group and one (11.1%) in the idiopathic group had concomitant pneumothorax. Tension pneumothorax appeared in a 5-year-old boy with severe pneumonia 2 months previous to this admission; he survived after intubation and chest tube insertion. A 9-month-old male infant had the longest hospital stay (53 days) due to bronchopneumonia concomitant with pneumothorax, PM and respiratory failure. A 15-year-old male with concomitant pneumothorax had recurrent pneumothorax for ~6 months afterward. He was tall and thin, and Marfan syndrome was highly suspected, because of other characteristic presentation. The infection group had higher incidence of pneumothorax and respiratory failure. All patients had complete or near complete resolution on chest radiography before discharge and were followed up as outpatients for 1–10 years. Hospital records showed no recurrence.

4. Discussion

The incidence of SPM in previous reports ranged from 1 in 800 to 1 in 42,000 patients in emergency

departments.⁸ Yellin et al screened routinely for SPM in young patients with chest pain and idiopathic dyspnea, and reported an incidence of 1 in 14,000. The bimodal peak in incidence represented children younger than 7 years and adolescents aged 13–17 years.¹¹ At Changhua Christian Hospital, another medical center in the center of Taiwan, the incidence was 1 in 15,150.⁷ In our series, the incidence was 1 in 8,302 at the Pediatric Emergency Department. Our age distribution also had a bimodal peak of children aged less than 4 years and adolescents aged 15–18 years.

Although SPM may appear spontaneously in children, trigger factors can be found in 70–90% of cases.^{3,9,10} In the present study, trigger factors were found in 50.0% of children with SPM, and infectious diseases was the main cause. Lower respiratory tract infections, such as bronchopneumonia and bronchiolitis, were the key trigger factors. There was neither vigorous chest physical therapy before SPM nor exacerbated PM after chest physical therapy. Two girls that presented with expiratory wheezing due to concomitant infection developed recurrent wheezing and asthma during the follow-up. It is likely they manifested SPM in their first asthma attack. Similar findings were also reported by Francesca et al.¹⁹ PM is a well-known complication in children with asthma and associated respiratory disorders.¹³ Stack and Caputo reported that 0.3% of 12,000 asthmatic children presented with SPM at a pediatric emergency department.^{2,11} Eggleston et al analyzed 479 patients aged 1–20 years hospitalized for asthma and found a mean SPM incidence of 5%.¹² Dekel et al followed patients and found they might have subclinical or clinical asthma according to clinical presentation and pulmonary function tests.³ In northern Taiwan, Chiu et al reported that asthma (50%) was the most common underlying factor.¹⁹ Because of the high prevalence of asthma related to SPM, diagnostic pulmonary function tests should be performed after the acute episode to confirm whether the child has asthma.

In our study, an 18-year-old boy was diagnosed during diabetic ketoacidosis (DKA). He presented with multiple episodes of strenuous vomiting with

Table 3 Hospital course of spontaneous pneumomediastinum in children

Cause	Associated pneumothorax, <i>n</i> (%)	ICU admission (days), <i>n</i>	ICU admission (days), mean \pm SD	Hospital length of stay (days), mean \pm SD
Infection	2 (25.0%)	7	9.1 \pm 12.3	13.0 \pm 14.0
Idiopathic	1 (11.1%)	4	1.5 \pm 0.87	4.0 \pm 2.6
Total	3 (16.6%)	11	6.4 \pm 10.0	7.9 \pm 11.5

ICU = intensive care unit; SD = standard deviation.

fresh blood, Kussmaul respirations and chest pain. Gastroscopy also revealed a Mallory-Weiss tear and hiatal hernia. The PM resolved spontaneously after starting DKA treatment. Cases of DKA in association with SPM have been reported.¹⁸ However, the exact pathophysiology of PM in association with DKA remains to be elucidated. Several hypotheses involve pressure gradient changes in the lung secondary to vomiting with subsequent increase in intrathoracic pressure.¹⁶ Kussmaul respiration can increase alveolar pressure by 20–30 mmHg, possibly sufficient to cause alveolar rupture.¹⁷ Donald et al questioned these hypotheses,¹⁸ observing chest pain before tachypnea and hyperemesis, and implied that PM sometimes precedes the onset of DKA, and perhaps initiates or hastens the progression of the metabolic abnormality. The prognosis of patients with SPM and DKA was excellent. Primary therapeutic attention to DKA results in an uneventful and rapid recovery.

In our study, idiopathic SPM occurred in nine patients (9/18, 50.0%), with mean age 12.1 ± 4.3 years, a rate far higher than in other studies (2–5%).^{3,13,14} All were boys, and six (6/9, 66.7%) with idiopathic SPM which developed after strenuous exercise before the attack of chest pain or dyspnea. These six patients presented with chest pain, and four also had dyspnea. One patient reported that the chest pain began after forcibly holding his breath when playing baseball. Clinical symptoms and signs subsided quicker than other groups, and their clinical courses were smooth. Although four patients (44.4%) required pediatric ICU admission (1.5 ± 0.9 days), most were discharged within 4 days (3.3 ± 1.7 days) and had an excellent outcome. In the infectious disease group, seven patients (87.6%) needed longer pediatric ICU admission (9.1 ± 12.3 days, $p=0.0197$) and hospitalization (13.0 ± 16.0 days, $p=0.0156$). The clinical course of idiopathic SPM in our study was similar to those in adult patients reported by other authors,^{9,10} who concluded that such patients might not need hospitalization or further specialized diagnostic study.

Esophageal rupture is extremely rare in children, but is a vital differential diagnosis for SPM.⁵ Such rupture may occur secondary to forceful vomiting or esophageal foreign body aspiration.²¹ A lateral decubitus radiography or esophagography with water-soluble contrast media would help preclude esophageal rupture. Foreign body aspiration was an unlikely cause of PM, with or without emphysema. Symptoms due to foreign body aspiration may mimic asthma and respiratory tract infection. Foreign bodies may penetrate the airway lumen and also cause non-penetrating airway obstruction, increasing the intra-alveolar pressure and leading to alveolar rupture.²² While no foreign body

aspiration- or esophageal rupture-related SPM occurred in this study, it is essential to rule out these factors in the initial differential diagnosis. Surgical or endoscopic intervention should be performed to prevent exacerbation.

The recurrence of SPM is quite low, with none in the present study. Most studies of SPM have reported no recurrence. Only a few case reports revealed recurrent SPM, in two patients with vomiting, two during asthma exacerbation, one during DKA, and one after athletic activity.^{23–25} Predisposing factors or underlying diseases should be labeled as risk factors and treated appropriately. Rest and avoidance of maneuvers that induce forced expiration should be advised to patients.

5. Conclusions

In conclusion, the incidence of SPM peaked in children younger than 4 years old and in those aged 15–18 years in central Taiwan. In toddlers and preschool children, infectious disease was the key trigger factor, without a sex difference. In adolescents, idiopathic factors, occasionally following agitated exercise, played an important role, particularly in males. SPM is a rare and usually benign condition that often resolves itself without sequelae. It is probably under-diagnosed in patients with complaints of chest pain, dyspnea and neck discomfort. While SPM is benign in itself, a major concern is the risk of commitment pneumothorax, pseudotamponade, underlying asthma, esophageal perforation and foreign body aspiration. Diagnosis is based on detailed physical examination and chest radiography. Hospitalization is required for evaluation and management of possible complications. Pulmonary function tests may be desired in children with SPM to detect the tendency for hyperactive airway. Foreign body aspiration is a rare cause, but pediatric clinicians should be alert for PM, which cannot be resolved without removing the foreign body.

References

1. Clalumeau M, Clainche LL, Sayeg N, et al. Spontaneous pneumomediastinum in children. *Pediatr Pulmonol* 2001;31:67–75.
2. Damore DT, Dayan PS. Medical causes of mediastinum in children. *Clin Pediatr* 2001;40:87–91.
3. Dekel B, Paret G, Szeinberg A, Vardi A, Barzilay Z. Spontaneous pneumomediastinum in children: clinical and natural history. *Eur J Pediatr* 1996;153:695–7.
4. Pierson DJ. Pneumomediastinum. In: Murray JF, Nadel JA, eds. *Textbook of Respiratory Medicine*, 2nd ed. Philadelphia: W.B. Saunders Co.; 1994:2250–65.
5. Rogers LF, Puig AW, Dooley BN, Cuello L. Diagnostic considerations in mediastinal emphysema: a pathophysiologic-roentgenologic approach to Boerhaave's syndrome and spontaneous pneumomediastinum. *AJR* 1972;115:495–551.

6. Hamman L. Spontaneous mediastinal emphysema. *Bull Johns Hopkins Hosp* 1939;64:1–21.
7. Han-Ping Wu, Ching-Yuang Lin, Chin-Fu Chang. Clinical spectrum of spontaneous pneumomediastinum in children. *J Pediatr Pulmo* 2004;02:96–104.
8. Nessian VJ. Recurrent pneumomediastinum in diabetic ketoacidosis. *Postgrad Med J* 1874;55:139–40.
9. Panacek EA, Singer AJ, Sherman BW, Prescott A, Rutherford WF. Spontaneous pneumomediastinum: clinical and natural history. *Am Emerg Med* 1992;21:51222–7.
10. Abolnik I, Lossos IS, Breuer R. Spontaneous pneumomediastinum: a report of 25 cases. *Chest* 1991;100:93–5.
11. Stack AM, Caputo GL. Pneumomediastinum in children asthma. *Pediatr Emerg Care* 1996;12:98–101.
12. Eggleston PA, Ward BH, Pierson WE, Bierman CW. Radiographic abnormalities in acute asthma in children. *Pediatrics* 1974;54:442–9.
13. Versteegh FGA, Broeders IAJM. Spontaneous pneumomediastinum in children. *Eur J Pediatr* 1991;150:304–7.
14. Grandibait B, Preminger A, Hatio B, Babyn P, Maayan C, Meizabav M. Pneumomediastinum and subcutaneous emphysema in a 18-month-old child. *J Pediatr* 2002;141:116–20.
15. Chalumeau M, Le Clainche L. Spontaneous pneumomediastinum in children. *Pediatr Pulmo* 2001;31:67–75.
16. Pooyan P, Puruckherr M, Summers JA, et al. Pneumomediastinum, pneumopericardium and peridural pneumatosis in DKA. *J Diabetes Complicat* 2004;18:242–7.
17. Bullaboy CA, Jennings RB Jr, Johnson DH, Coulson JD, Young LW, Wood BP. Radiological case of the month. Pneumomediastinum and subcutaneous emphysema cases by diabetic hyperpnea. *Am J Dis Child* 1989;143:93–4.
18. Donald E Girard, Victor Carlson, Ethan A Natelson, Herbert L Fred. Pneumomediastinum in diabetic ketoacidosis: comments on mechanism, incidence and management. *Chest* 1971;60:455–9. AQ6
19. Chiu CY, Wong KS, Tao TS, Huang JL. Asthmatic versus non-asthmatic spontaneous pneumomediastinum in children. *Asian Pac J Allergy Immunol* 2005;23:19–22.
20. Bullaro Fm, Bartoletti SC. Spontaneous pneumomediastinum in children: a literature review. *Pediatr Emerg Care* 2007; 23:28–30. AQ7
21. Engum SA, Grosfeld JL, West KW, Roscola FJ, Tres Scherer LR, Vaughan WG. Improved survival in children with esophageal perforation. *Arch Surg* 1996;131:604–10.
22. Burton EM, Riggs W, Kaufman RA, Houston CS. Pneumomediastinum caused by foreign body aspiration in children. *Pediatr Radiol* 1989;20:45–7.
23. Bullimore DW, Cooke D. Cyclical vomiting with pneumomediastinum. *Acta Pediatr Scand* 1982;71:675–6.
24. Channer KS, Roberts CJC, Jeyasingham K. Recurrent idiopathic pneumomediastinum. *Postgrad Med J* 1985;61:1073–5.
25. Nessim VJ. Recurrent pneumomediastinum in diabetic ketoacidosis. *Postgrad Med J* 1974;55:139–40.

Author Queries

AQ1: Please check the use of SPM and PM abbreviations are correct throughout the manuscript.

AQ2: posteroanterior?

AQ3: anteroposterior

AQ4: Please confirm the image used for Figure 1 as there were 2 similar figures provided

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