

# Etiologies of Spontaneous Pneumomediastinum in Children in Middle Taiwan

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**Summary.** Introduction: Spontaneous pneumomediastinum (SPM), while rare, is probably underestimated in children. Treatment targets on the underlying disease and trigger factors. The study aimed in analysis different etiology in different age groups. Patients and Methods: Total 37 children with SPM were analyzed from two medical centers in middle Taiwan from 1994 to 2007. Results: Incidence of SPM in children was 1:11,726 patients at Department of Pediatric Emergency in middle Taiwan. Bimodal peak in incidence occurred in those under 7 and in those aged 15–18 years old. The Characteristic symptoms were dyspnea (64.9%), followed by chest pain (62.2%) and neck pain (40.5%); common specific physical signs were subcutaneous emphysema (SCE) (67.6%) and Hammer's sign (13.5%). Trigger factors were infection (43.2%), asthma (21.0%), esophageal rupture (5.4%), foreign body aspiration (2.7%), and diabetic ketoacidosis (2.7%). Idiopathic SPM accounted for 35.1% of patients with mean age 14.1 years. In age distribution, preschoolers (<7 years old) got SPM mostly due to lower respiratory tract infection. In adolescents, the most common etiologies were asthma and upper respiratory tract infection. Mean hospitalization was 6.4 days. Although 17 (46.0%) patients needed intensive care, nearly all had complete resolution in chest radiography before discharge. Conclusion: Clinician should keep alert to incidence of SPM from these symptoms. Etiologies varied with age and treatment must target on factors and underlying disease. *Pediatr Pulmonol.* 2010; 45:869–873. © 2010 Wiley-Liss, Inc.

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## INTRODUCTION

Spontaneous pneumomediastinum (SPM) is preserved for patients of mediastinal air leak without chest trauma, mechanical ventilator, iatrogenic procedure or surgery. SPM can occur due to air leakage from intra- or extrathoracic source<sup>1,2</sup> but rarely before age 18.<sup>1,3</sup> In most cases, air leakage from ruptured alveolar escapes and dissects the hilum along peribronchovascular sheaths and spreads to mediastinum.<sup>4,5</sup> Once in the mediastinum, air extends around the large vessel and esophagus to thoracic wall. From chest radiographical signs indicated pneumomediastinum: for example, vertical lucent streak along the left side of heart and aortic arch, continuous diaphragm sign, lucent streak along retrosternal, pericardiac and peritracheal area, logical according to pathogenesis of PM. Yet SPM was diagnosed by chance on chest radiography in a patient with isolated chest pain. Children with SPM might present chest pain, subcutaneous emphysema (SCE), dyspnea and Hamman's sign. Rarely, tension pneumomediastinum leads to airway compression and venous obstruction.

SPM was first described by Hamman in 1939<sup>6</sup> and became increasingly recognized as a distinct clinical entity. Most articles about PM have been conducted in

adults and few studies in children. Possible precipitating or trigger factors have been reported in 70–90% of cases from previous studies.<sup>3,7,8</sup> Respiratory maneuver and Valsalva maneuver, such as coughing, crying-screaming, vomiting, hypercapnea, competitive athletics, illicit drug inhalation, etc., were cited as causes in children. Medical conditions like asthma, bronchiolitis, bronchopneumonia, convulsion or laryngitis, were also linked.<sup>1</sup> This study aimed at character of clinical presentation, precipitating factors, hospital course, and possible sequel in children of different ages.

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## PATIENTS AND METHODS

### Patients Population

Retrospective chart review of 25 PM patients aged 6 months to 18 years was conducted at China Medical University Hospital (CMUH) from November 1997 to July 2007 according to 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, trauma, iatrogenic procedure, surgery or with history of underlying lung disease were excluded from further analysis. Eighteen patients were monitored at CMUH. Besides, these data were combined with 19 patients collected under similar criteria from Changhua Christian Hospital from June 1994 to June 2003.<sup>9</sup> A total 36 patients of SPM were further studied in the series.

### Methods

Diagnosis was made from PA view or AP view of chest radiography, including cervical region. adding lateral chest films improved diagnostic accuracy considerably. 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. Review of medical records and history focused on identifying predisposition or trigger factors like infectious disease, medical condition or strenuous sport 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, SCE or Hammer's sign. Hammer's sign is defined as a crunching noise, made by air in mediastinal tissues, almost pathognomonic for this condition and at times linked with reduction in heart sound. Previously healthy children developing SPM without apparent predisposing or trigger factors were classified in the idiopathic group. For suspected esophageal rupture, esophagography was arranged. Severity could be related to the volume of escaping air into mediastinum, airway compression with respiratory distress and obstruction of venous return with pseudotamponade. Intensive care should be considered under impending decompensation. Length of stay and the incidence of intensive care unit (ICU) care were further analyzed.

## RESULTS

### Characteristics of Patients With SPM

Mean age of 28 boys (75.7%) and 9 girls (24.3%) was 10.3 years old with a range of 9 months to 18 years. None

**TABLE 1—Clinical Causes and Demographics of Spontaneous Pneumomediastinum in Children**

	No (%)	Age (year) mean	Gender (M/F)
Infections	16 (43.2)	6.7	10:6
Bronchopneumonia	7 (19.0)	6.4	4:3
Bronchiolitis	4 (10.8)	2.5	3:1
URI	3 (8.1)	11.3	2:1
Laryngitis	1 (2.7)	18.0	1:0
Croup	1 (2.7)	1.0	0:1
Asthma	4 (10.8)	15.8	4:0
Esophageal rupture	2 (5.4)	3.0	1:1
Foreign body aspiration	1 (2.7)	2.3	0:1
DKA	1 (2.7)	18	13:1
Idiopathic	13 (35.1)	14.1	1:0
Total	37 (100)	10.3	28:9

URI, upper respiratory infection; DKA, diabetic ketoacidosis.

smoked or used drugs. The presence of SPM was confirmed radiographically in all patients. Bimodal peaks in incidence occurred under 7 years and from 15 to 18 years old. Boys were predominant.

### Etiology

Table 1 summarizes the clinical course and demographics. Most common cause in this series was infection (16/37, 43.2%). Thirteen patients (13/37, 35.1%) were identified as idiopathic group. One patient had SPM complicated with diabetic ketoacidosis (DKA) (1/37, 2.7%). One patient (1/37, 2.7%) had SPM due to foreign body aspiration, and two patients (2/37, 5.4%) due to esophageal rupture. In the infectious disease group, bronchopneumonia and bronchiolitis were two leading causes. Most had wheezing in breath sound from physical examination; two (5.4%) girls developed asthma later—that is, manifested SPM in the first asthma attack. As for age distribution, infection was the leading cause of SPM in preschool age and middle childhood (mean: 6.7 years old). In infectious disease, seven patients (7/16, 43.8%) had bronchopneumonia with mean age 6.4 years, four patients (4/16, 25%) had bronchiolitis with mean age 2.5 years and three patients (3/7, 42.9%) had upper respiratory tract infection with mean age 11.3 years. One 1-year-old girl had SPM due to croup and one 18-year-old boy due to laryngitis. In preschoolers (<7 years old), lower respiratory tract infection was the major cause of SPM. In adolescents ( $\geq 10$  years old), asthma and upper respiratory tract infection were predominant but idiopathic SPM was more common. Six (6/13, 46.2%) of all male adolescents had idiopathic SPM after strenuous exercise.

### Clinical Findings

Table 2 plots associated presenting symptoms and signs, predominantly dyspnea and chest pain. The latter

**TABLE 2—Clinical Presentation of Spontaneous Pneumomediastinum in 37 Children**

Symptoms	No. (%)	Signs	No. (%)
Dyspnea	24 (64.9)	SCE	25 (67.6)
Chest pain	23 (62.2)	Hamman's sign	6 (16.2)
Neck pain or sore throat	21 (56.8)	Neither	8 (21.6)
Dysphagia	4 (10.8)	Both	3 (8.1)
Back pain	4 (10.8)		
Shoulder pain	2 (5.4)		

SCE, subcutaneous emphysema.

worsened with deep respiration and postural changing in seven patients (16/37, 43.2%) and radiated to back, shoulder or arms in seven patients (7/37, 18.9%). Most common clinical sign was SCE (67.6%). Five (13.5%) patients had Hamman's sign, three (8.1%) both SCE and Hamman's sign. Eight patients (21.6%) had neither SCE nor Hamman's sign.

### Hospital Courses

Table 3 reveals hospital courses. All patients were admitted to the hospital, 17 (45.9%) requiring intensive care due to respiratory distress. Nine patients had infectious disease, one asthma attack, one foreign body aspiration, one esophageal rupture and four idiopathic SPM. Risk factors for intensive care were young age and infection. Length of stay in hospital was 6.4 days with a range of 2–53 days. Idiopathic group had hospitalization 3.5 days and infection group 8.8 days. A 9-month-old male infant had longest hospital stay (53 days) due to bronchopneumonia concomitant with pneumothorax, pneumomediastinum and respiratory failure.

### Treatment and Outcome

Main treatments were primary analgesics and management of underlying condition. Chest pain was relieved by acetaminophen and/or nonsteroid anti-inflammatory drugs (NSAID). Bronchodilator or prokinetic agents was

**TABLE 3—Hospital Courses of Spontaneous Pneumomediastinum in Children**

Causes	Associated pneumothorax	ICU admission (n)	Hospital length of stay (mean, days)
Infections	5	9	9.0
Asthma	3	1	4.2
Foreign body aspiration	1	1	6.0
Esophageal rupture	1	2	8.5
Idiopathic	3	4	3.0
Total	13	17	6.3

prescribed for apparent wheezing, asthma attack, and/or vigorous vomiting. Rest and oxygen therapy were requested in all patients to prevent progression to pneumothorax and respiratory distress.

### Complication

Thirteen patients (13/37, 35.1%) had concomitant pneumomediastinum, pneumothorax and dyspnea; three received chest tube insertion, two endotracheal tube insertion and ventilator. Five patients had concomitant pneumothorax (5/13, 38.7%) due to infection and three (3/13, 23.1%) was idiopathic. From different etiologies, foreign aspiration (1/1), asthma (3/4) and esophageal rupture (1/2) had higher potential for developing concomitant pneumothorax. None of them developed tension pneumothorax and they resolved gradually without thoracostomy. Tension pneumothorax appeared in a 5-year-old boy with severe pneumonia 2 months ago; he survived after intubation and chest tube insertion. A 9-month-old male infant had longest hospital stay (53 days) due to bronchopneumonia concomitant with pneumothorax, pneumomediastinum and respiratory failure. A 15-year-old male with concomitant pneumothorax had recurrent pneumothorax for half a year afterward. He was tall and thin, with Marfan syndrome highly suspected, according to other characteristic presentation. Infection had higher incidence of pneumothorax and respiratory failure. All patients had complete or nearly complete resolution on chest radiography before discharge and were followed up as outpatients for 1–10 years. Hospital records showed no recurrence.

### DISCUSSION

Incidence of SPM in previous reports ranged from 1 in 800 to 1 in 42,000 patients in emergent department.<sup>1</sup> Yellin et al. screened routinely for SMP in young patients with chest pain and idiopathic dyspnea and found an incidence of 1 in 14,000. Bimodal peak in incidence was younger than 7 years and in adolescents aged between 13 and 17 years.<sup>10</sup> In our series, incidence was 1 in 11,726 at the Pediatric Emergency Department at medical centers in middle Taiwan. Our age distribution also had a bimodal peak under 7 years and from 15 to 18 years.

Although SPM may appear spontaneously in children, trigger factors can be found in 70–90% of cases.<sup>3,7,8</sup> In the present study, trigger factors found in 64.9% of children with SPM and infectious diseases is the major reason. Among infectious diseases, lower respiratory tract infection such as bronchopneumonia and bronchiolitis were key trigger factors. There was neither vigorous chest physical therapy before SPM nor exacerbated PM after chest physical therapy. Two girls that presented expiratory wheezing due to concomitant infection developed recurrent wheezing and asthma in follow-up. They might

manifest SPM in their first asthma attack. Similar finding was also reported by Bullaro et al.<sup>11</sup> Pneumomediastinum was a known complication in children with asthma and associated respiratory disorders.<sup>12</sup> Stack and Caputo report 0.3% of 12,000 asthmatic children presenting SPM at a pediatric emergency department.<sup>13</sup> Eggleston et al.<sup>14</sup> analyzed 479 patients aged 1–20 years hospitalized for asthma and found a mean SPM incidence of 5%. Dekel et al.<sup>3</sup> followed patients and found they might have subclinical or clinical asthma according to clinical presentation and pulmonary function tests. In Northern Taiwan, Chiu et al.<sup>15</sup> reported asthma (50%) as the most common underlying factor.<sup>13</sup> Owing to 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 DKA. He presented multiple episodes of strenuous vomiting with fresh blood, Kussmaul respirations and chest pain. Gastroscopy also revealed Mallory-Weiss tear and hiatal hernia. PM resolved spontaneously after DKA treatment commenced. Cases associated with DKA and SPM have been reported.<sup>16</sup> The exact pathophysiology of PM in association with DKA remain to be elucidated. Several hypotheses involve pressure gradient changes in the lung secondary to vomiting with subsequent increase in intrathoracic pressure.<sup>17</sup> Kussmaul respiration can increase alveolar pressure by 20–30 mmHg, possibly sufficient to cause alveolar rupture.<sup>17,18</sup> Donald et al. questioned the hypotheses,<sup>17</sup> observing chest pain before hyperpnea and hyperemesis, then implied that pneumomediastinum sometimes precedes onset of DKA, perhaps initiates or hastens progression of the metabolic abnormality. Prognosis in patients with SPM and DKA was excellent. Primary therapeutic attention to DKA results in uneventful and rapid recovery.

We generally consider spontaneous pneumothorax (SPX) and pneumomediastinum (SPM) in children as showing different etiology, predisposition, clinical course and therapeutic management. Spontaneous pneumothorax most often occurs in patient at rest or with minimal exertion.<sup>19</sup> Air leaks mainly from peripheral alveolar and visceral pleural, then accumulates in pleural space. Structural abnormalities appear: for example, congenital or acquired bullae, bleb, or cyst. With pneumomediastinum, air leaks from the esophagus, the central alveolar or bronchial tree and then dissects along peribronchial and perivascular tissue plane centrifugally toward hilum or peripherally toward the visceral pleural surface, most often with relatively less structural abnormality or trigger factors. This leads to bronchial hyper-reactivity or barotraumas. Some articles reported no concurrent pneumothorax,<sup>3</sup> while one-third of our patients had both it and concomitant pneumomediastinum. Table 3 plots SPM associated with pneumothorax in infection and

esophageal rupture, plus longer hospitalization. It might mean overlapping mechanism between SPM and SPX.

Esophageal rupture is extremely rare in children but a vital differential diagnosis for SPM.<sup>5</sup> Such rupture may occur secondary to forceful vomiting or esophageal foreign body aspiration.<sup>20</sup> A lateral decubitus radiography or esophagography with water-soluble contrast media would help preclude esophageal rupture. Two younger (3.0 years old) patients had SPM due to esophageal rupture, both admitted to ICU and hospitalized longer (8.5 days) than average. Spontaneous esophageal rupture (effort rupture of esophagus or Boerhaave's syndrome) most often results from a sudden increase in intra-esophageal pressure combined with negative intrathoracic pressure due to straining or vomiting.<sup>21</sup> Classical triad of esophageal rupture includes vomiting, lower chest pain and cervical SCE. Dysphagia, dyspnea, pleural effusion, mediastinitis, and hemodynamic instability were possible complications, a life-threatening condition warranting early diagnosis and prompt attention. Esophageal evaluations such as contrast swallowing study, esophagogastroscopy or computed tomography should be performed on suspected cases.

Foreign body aspiration was an unlikely cause of pneumomediastinum, with or without emphysema. Symptoms of foreign body aspiration may mimic asthma and respiratory tract infection. Foreign bodies may penetrate airway lumen and also cause nonpenetrating airway obstruction, increasing intra-alveolar pressure and leads to alveolar rupture.<sup>22</sup> One foreign body aspiration and two esophageal rupture related SPM occurred in this study. All of them was admitted to ICU and had high rate of concomitant pneumothorax, 50% and 100% respectively. Although few case number, it is crucial to rule out these factors in initial differential diagnosis. Surgical or endoscopy intervention should be performed to prevent exacerbation.

In our study, idiopathic SPM occurred in 13 patients (13/37, 35.1%) with mean age 14.1 years, a rate far higher than in other studies (2–5%).<sup>3,12,23</sup> All were boys, six (6/13, 46.2%) with idiopathic SPM had strenuous exercise before the attack of chest pain or dyspnea. These six presented chest pain, and four also had dyspnea. One patient clearly recalled that chest pain began after forcibly holding breath when playing baseball. Clinical symptoms and signs subsided quicker than other groups, and their clinical courses were smooth. While four (30.8%) patients required pediatric ICU admission (1.5 days), most were discharged within 3 days (3.0 days) and had excellent outcome. While in infectious disease, nine (56.2%) patients needed pediatric ICU admission (56.3%) and longer hospitalization (8.8 days). The clinical courses of idiopathic SPM in our study were similar to those in adult patients reported by other authors<sup>7,8</sup> who concluded that the patient might not need hospitalization and further specialized diagnostic study.

Recurrence of SPM was quite low, none in the present study. Most SPM associated papers report no recurrence. Only few case reports revealed recurrent SPM in two patients with vomiting, two during asthma exacerbation, one during DKA, and one after athletic activity.<sup>24–26</sup> Predisposing factors or underlying diseases should be labeled risk factors and treated properly. Rest and avoidance maneuvers of creating forced expiration were reasonable to advise the patients.

In sum, incidence peaked in those younger than 4 years and in those from 15 to 18 years in middle Taiwan. In toddlers and preschool children, infectious disease was the key trigger factor without gender difference. In adolescents, idiopathic factor, occasionally following agitated exercise, played an important role, especially in males. SPM is a rare and usually a benign condition which often resolves itself without sequel. It is probably underdiagnosed in patients with complaints of chest pain, dyspnea, and neck discomfort. While SPM is benign in itself, a major concern is risk of commitment pneumothorax, pseudotamponade, underlying asthma, esophageal perforation and foreign body aspiration. Diagnosis was based on detail physical examination and chest radiography. Hospitalization is required for evaluation and management of possible complication. Pulmonary function test might be desired for children with SPM to detect tendency of hyperactive airway. Esophageal rupture and foreign body aspiration were rare causes, but pediatric clinicians should keep alert for pneumomediastinum and concomitant pneumothorax that cannot be resolved without removing the foreign body.

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