Spontaneous Pneumothorax in Children

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Objectives

• Definition and Classification
• Epidemiology
• Pathophysiology
• Clinical Presentation and Diagnosis
• Management
Pneumothorax

Collection of air in the pleural space (between the visceral and parietal pleura) separating lung from the chest wall.

Pneumothorax

- Traumatic
  - Spontaneous
    - Primary (PSP)
    - Secondary (SSP)
Epidemiology of PSP

• Adult data
  – Male preponderance
  – Men: 7.4 – 18 per 100,000 men
  – Women: 6 per 100,000
  – Peak incidence between 16 and 24 years of age

• Pediatric Data
  – Incidence unclear
  – Male predominance in adolescents but probably not in younger children
  – Peak incidence bimodal: most cases in neonates or late adolescence
  – Low incidence suggested by multiple retrospective studies


<table>
<thead>
<tr>
<th>Study</th>
<th>Institution</th>
<th>#PSP patients (# male)</th>
<th>Period of Study</th>
<th>Age (years; given as mean +/- SD)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Davis et al 1993</td>
<td>Royal Children's Australia</td>
<td>10 (6)</td>
<td>25 years</td>
<td>14.6 (range 11.8-17.4)</td>
</tr>
<tr>
<td>O'Late et al 2008</td>
<td>Royal Children's Australia</td>
<td>15</td>
<td>7 years</td>
<td>14.4 (range 1-18)**</td>
</tr>
<tr>
<td>Qureshi et al 2005</td>
<td>Children's at Pittsburgh</td>
<td>43 (35)</td>
<td>13 years</td>
<td>15.9 +/- 0.3</td>
</tr>
<tr>
<td>Cook et al 1999</td>
<td>Columbus Children's</td>
<td>15 (12)</td>
<td>5.5 years</td>
<td>14 +/- 1.1</td>
</tr>
<tr>
<td>Poenaru et al 1994</td>
<td>Hospital St. Justine, Montreal</td>
<td>38 (38)</td>
<td>20 years</td>
<td>15 (range 2-22)**</td>
</tr>
<tr>
<td>Wilcox et al 1995</td>
<td>Children's Hospital of Buffalo, NY</td>
<td>10 (11)</td>
<td>13.5 years</td>
<td>13.8 (range 2-16)**</td>
</tr>
<tr>
<td>Shaw et al 2003</td>
<td>Montreal Children's Hospital</td>
<td>50 (not given)</td>
<td>10 years</td>
<td>Not given</td>
</tr>
</tbody>
</table>

**Includes cases of secondary pneumothorax

Robinson et al; “Evidence-based Management of paediatric primary spontaneous pneumothorax”, Paediatric Resp Reviews 10 (2009) 110-117 (Adapted from Table 2)
Risk Factors

- Patients typically are tall with thin body habitus
- Smoking
  - Healthy adult male smokers 12% risk vs. 0.1% risk in non-smoking men
- Other precipitating events: military flying, weight lifting, diving, Valsalva maneuver
- Familial cases are described; usually adult or adolescent males

Secondary Pneumothorax (SSP)

<table>
<thead>
<tr>
<th>Classification</th>
<th>Disease</th>
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<tbody>
<tr>
<td>Airway Disease</td>
<td>Cystic Fibrosis, Asthma, COPD</td>
</tr>
<tr>
<td>Infection</td>
<td>Pneumocystis carinii, TB, Necrotizing pneumonia</td>
</tr>
<tr>
<td>Congenital/Neonatal</td>
<td>Congenital pulmonary adenomatous malformation (CCAM), Congenital lobar emphysema</td>
</tr>
<tr>
<td>Interstitial</td>
<td>Sarcoidosis, Langerhans’-cell granulomatosis</td>
</tr>
<tr>
<td>Connective Tissue or Inflammatory</td>
<td>Marfan syndrome, Ehlers-Danlos, Juvenile idiopathic arthritis, polymyositis or dermatomyositis</td>
</tr>
<tr>
<td>Malignancy</td>
<td>Primary lung cancer, metastatic</td>
</tr>
<tr>
<td>Airway Obstruction</td>
<td>Foreign body aspiration</td>
</tr>
<tr>
<td>Catamenial</td>
<td>PTX triggered by menstrual cycle, thoracic endometriosis</td>
</tr>
</tbody>
</table>
Pathophysiology

The Pleural Space

Visceral Pleura
- Covers the lung parenchyma
- Provides mechanical support to the lung
- Limits lung expansion, protecting the lung
- Contributes to the elastic recoil of the lung and lung deflation.

Parietal Pleura
- Lines the inside of the thoracic cavities.
- Subdivided into the costal, mediastinal and diaphragmatic parietal pleura.
- Loose connective tissue and single layer of mesothelial cells.

Pathophysiology

Pleural Pressure

- Negative pressure generated between the visceral and parietal pleura by the opposing elastic forces of the chest wall and lung at Functional Residual Capacity (FRC)
  - FRC is volume of air present in the lungs at the end of passive expiration.

Pleural pressure represents the balance between the outward pull of the thoracic cavity and the inward pull of the lung.
Pleural Pressure at FRC

- Uncouples lung and chest wall
  - Air introduced to pleural space → raising intrapleural pressure to atmospheric
- Lungs and chest wall move to their unstressed volumes:
  - Lungs recoil inward
  - Chest wall recoils outward

Pneumothorax
Effects of Pneumothorax on Pleural Pressure

Air will flow into the pleural space until a pressure gradient no longer exits or until the communication is sealed.

Pathogenesis PSP

Pathophysiologic mechanisms are poorly understood

Large increases in transpulmonary pressure

- Transpulmonary pressure: $P_{alveolar} - P_{pleural}$
- Alveolar distension $\rightarrow$ alveolar rupture
- Can get rapid large shifts in pressure with spontaneous respiratory efforts, positive pressure ventilation, Valsalva, or airway-obstruction (creating ball-valve effect)
Pathogenesis of PSP

- Sub-pleural blebs and bullae are found at lung apices in majority of PSP patients
  - 56-88% of adult PSP patients
  - 2 small observational pediatric studies: 45% (5 of 11 CT scans) and 100% (3 of 3 CT scans)

• Unclear how often lesions are site of air leak
  – Only a majority are ruptured at time of thoracoscopy

Pathogenesis of PSP

Defects in Visceral Pleura

- Increase in pleural porosity secondary to inflammation
- Areas of disrupted mesothelial cells at the visceral pleura → replaced by inflammatory elastofibrotic layer with increased porosity allowing air leakage into pleural space
- Blebs/bullae and areas of pleural porosity may be linked to variety of factors:
  – Distal airway inflammation
  – Hereditary predisposition
  – Anatomical abnormalities in bronchial tree
  – Abnormal connective tissue
- Direct injury due to underlying lung disease (SSP)
  – Local inflammation and tissue necrosis → bronchopleural fistula
  – May explain pneumothorax associated with necrotizing pneumonia, TB, abscess, CF, connective tissue disease (Marfan), etc.
Clinical Presentation

- Most common: acute onset chest pain and dyspnea
  - Diffuse on affected side, radiates to ipsilateral shoulder
- Usually occurs at rest
- May be precipitated by maneuvers that increase intrathoracic pressure (valsalva – lifting/straining)

<table>
<thead>
<tr>
<th>Size/Type</th>
<th>Clinical Findings</th>
</tr>
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<tbody>
<tr>
<td>Small</td>
<td>Tachycardia (most common), dyspnea</td>
</tr>
<tr>
<td>Large</td>
<td>Ipsilateral decrease/absent breath sounds, hyper-resonant percussion note, ipsilateral decreased vocal fremitus, tachypnea, WOB, cyanosis</td>
</tr>
<tr>
<td>Tension</td>
<td>Tracheal deviation to contralateral side, tachycardia, hypotension, shock, cyanosis, diminished heart sounds</td>
</tr>
</tbody>
</table>

Diagnosis

Source: wikiradiography.net
Diagnosis
Chest CT

• Can be used to detect small PTXs
• Provide more detailed information to assist in subsequent management
  – Number, size, location of bullae/blebs

• More than 90% of adult patients with PSP, pathological changes can be detected on chest CT
• Frequency of blebs in children with PSP varies widely (range 28-45%)
• Unclear clinical significance of apical blebs < 2 cm
• Several studies suggest presence of apical blebs on CT does not predict future recurrence risk

Source: ctsnet.org
Estimation of Size

**Adults**
- Large: >3cm air b/w pleural line and chest wall OR >2 cm b/w entire lateral lung edge and chest wall
- Correspond to ~20-30% PTX
- Accuracy limited
- *Size of PTX is less important than the degree of clinical compromise in defining management strategy*

**Children**
- No standard methods
- Use of adult methods in adolescents likely reasonable
- Younger children: “small” or “large” based on relative size of PTX compared with thoracic cavity

Treatment

- No standardized guidelines for children
- British Thoracic Society (2010) and American College of Chest Physicians have published guidelines for adults
- Paucity of evidence to guide management illustrated by lack of consensus between different adult guidelines
- Extrapolate adult guidelines to children
- *Caution: need to individualize approach with careful monitoring*
  - *Estimates of pneumothorax size and clinical outcomes are not well-defined in children*
Treatment Approaches in Children

Stable, 1st small PSP
- Observe in hospital
- Supplemental O2
- Serial CXRs

Conservative Management
- If PSP is small and decreasing in 12h
- Continued observation as outpatient
- Younger patients (<12 y): observe longer

Supplemental Oxygen
- 100% FIO2 via NRB
- Hastens reabsorption of intra-pleural air
- Limited evidence from small studies
Treatment Approaches in Children

Large PTX (>3cm rim of air)
dyspnea, hypoxemia

Supplemental Oxygen
Needle aspiration
If unstable → chest tube or pigtail
SSP → chest tube or pigtail

Needle Aspiration

- Large bore IV + syringe + 3 way stopcock
- Withdraw air until stops
- Continuous air = air leak → chest tube
- If no leak, close stopcock, secure catheter
- Observe 4 hours, then CXR
  - If PTX evacuated, d/c catheter and observe 24 hours with close monitoring and telemetry
  - Follow Serial CXRs
    - Air will re-accumulate in 20-50% due to air leak
    - Repeat CXR prior to discharge
    - If recurrent PTX → chest tube
Thoracostomy or Catheter Evacuation

- Failed aspiration, recurrent PSP, SSP
- Pleur-Evac to water seal to prevent re-accumulation of air
- If lung not fully re-expanded with drainage → place to suction
  - Early use of strong suction not recommended due to risk of re-expansion induced pulmonary edema
- If not air leak after 12 hours (bubbles), clamp CT for ~24 hours (utility is an unproven approach) followed by CXR
- Remove CT if no recurrent PTX

Thoracostomy Tube Pigtail Catheters
Surgical Intervention

• Controversial
  – Unclear if surgery will prevent recurrence

• Pleurodesis
  – Chemical (talc, tetracycline, fibrin glue) done at time of CT placement
  – Stimulates inflammatory response → adherence of visceral and parietal pleural surfaces
  – Mechanical abrasion- more common in children

• Stapling/oversewing ruptured blebs or tears in visceral pleura and resection of abnormal lung tissue (blebectomy or bullectomy), parietal pleurectomy
  – Performed via VATS or mini or conventional thoracotomy
  – Pleurodesis often performed with surgery

Proposed Indications for Surgical Intervention

• 1st PSP with persistent air leak
  – Timing debatable: ACCP guidelines = 4 days, BTS guidelines 5-7 days

• 2nd ipsilateral PSP

• 1st contralateral PSP

• Bilateral pneumothorax

• Spontaneous hemothorax

• SSP – depends on type and severity of underlying lung disease
  – Individualized treatment decisions
  – Recurrent SSP
  – CF and recurrence of large SSP
Pediatric Literature

- No evidence to support use of CT scans and subsequent surgical intervention in 1st episodes of PSP
- Recent case series (Qureshi et al J Pediatr Surg 2005) compared VATS at initial episode vs. VATS at 1st recurrence
  - Increase rate of recurrence in initial VATS group (29% vs. 0%)

Robinson et al., "Management of paediatric spontaneous pneumothorax"; Arch Dis Child 2015

- Multicentre retrospective review, presenting to EDs in Australia and New Zealand
- Comparing PSP and SSP management: conservative, ICC, aspiration, later surgery
- 162 children, 219 episodes of spontaneous PTX
- Median age 15 years, 71% male

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<thead>
<tr>
<th></th>
<th>PSP</th>
<th>SSP</th>
<th>p value</th>
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<tbody>
<tr>
<td># episodes</td>
<td>155</td>
<td>64</td>
<td></td>
</tr>
<tr>
<td>Active intervention (%)</td>
<td>85/155 (55%)</td>
<td>45/64 (70%)</td>
<td>&lt;0.05</td>
</tr>
<tr>
<td>ICC as 1st line</td>
<td>63/86 (73%)</td>
<td>41/45 (91%)</td>
<td>&lt;0.05</td>
</tr>
<tr>
<td>Initial aspiration</td>
<td>23/86 (27%)</td>
<td>4/45 (9%)</td>
<td>NS</td>
</tr>
<tr>
<td>ICC as any intervention</td>
<td>74/155 (48%)</td>
<td>43/64 (67%)</td>
<td>&lt;0.01</td>
</tr>
</tbody>
</table>
Robinson et al.

- 42% received conservative management
  - 46% discharged home from ED
- Aspiration success 52%
  - No difference b/w PSP and SSP
  - Greater success in smaller vs. larger PTX
  - Overall recurrence rate 95%
    - Median interval b/w presentations was 62 days

Summary
- Management differs from adult guidelines
- For PSP, ICC is 1st line intervention
- Compared to PSP, episodes of SSP have higher rate of active intervention, ICC insertion and longer duration of admission
- Aspiration use low, did not differ b/w PSP and SSP
- Clinicians tend not to differentiate b/w PSP and SSP
- Need large observational study to clarify optimal management strategies in children

O’Lone et al.; "Spontaneous Pneumothorax in Children: When is Invasive Treatment Indicated?"; Ped Pulm 2008

- Retrospective review Royal Children’s Hospital, Melbourne Australia 1997-2004
- 31 patients: 15 PSP, 16 SSP
  - 7 patients with previous PTX
- Mean age 14.4 years

Fig. 1. Primary Treatment of spontaneous pneumothorax (n=35)
O’Lone et al.; “Spontaneous Pneumothorax in Children: When is Invasive Treatment Indicated?”; Ped Pulm 2008

- 25 episodes (69%), including 14/22 treated with ICC resolved with conservative therapy
- Mean ICC days = 4.9 (range 2-10 days)
- 25 patients with 1st PTX:
  - 9 treated successfully with observation
  - 16 with ICC
  - 4/16 required surgical intervention after ICC
- 8 patients with ICC < 5 days
  - 1 had recurrence requiring surgical treatment on 2nd admission
  - 1 had apical bullus
  - 6 had no problems

- 6 patients with ICC > 5 days
  - 1 (CF) died
  - 1 recurrence
  - 4 found to have bullae on CT
- 11 episodes (30.5%) required secondary treatment during same admission with air leak avg 5.9 days
  - 2 CF, 3 Marfan
  - 9 required invasive procedure
- 11 follow up studies: 45% had apical abnormalities on CT
**Conclusions**

- Higher incidence of recurrence and underlying abnormalities in patients whose air leak persisted > 5 days
- Decision to progress to invasive procedure dependent on individual patient characteristics and underlying disease
- Consider early invasive treatment (within 48h) for recurrent PTX
- Recommend invasive therapy if air leak > 5 days
  - In true PSP, resolution may occur with up to 10 days of conservative therapy
  - However, in all patients with air leak > 5 days, an underlying surgically resectable structural cause found

**Prognosis**

**A few case series**

- 21% recurrence in 171 adolescents with PSP (2 year follow-up)
  - Large PTX with persistent air leak influenced recurrence
  - VATS reduced risk of ipsilateral recurrence (4% with VATS vs. 18% without VATS)
  - VATS group had 18% contralateral recurrence
- 51% recurrence among 58 children with PSP managed conservatively
- 55% ipsilateral and 15% contralateral recurrence among 27 patients with PSP whose leak initially closed during non-operative management
  - 20 patients with persistent air leak had VATS or thoracotomy: 15% recurrence
  - CT not helpful in predicting recurrence on contralateral side
References

- Janahi, I; "Spontaneous pneumothorax in children"; UpToDate 2015.