Congenital pulmonary malformations and lung cancer

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The continuum spectrum of Bronchopulmonary Congenital Anomalies

FOREGUT ANOMALIES

Bronchogenic Cyst
Enteric duplication
Neuroenteric cyst
TEF/Diverticula Stenosis

Pulmonary Sequestration
Pulmonary agenesis / Hypoplasia
CLE - CCAM

Bronchial atresia
Tracheal atresia
Tracheal bronchus

PULMONARY PARENCHYMA ANOMALIES

LUNG PARENCHYMA ANOMALIES

VASCULAR ANOMALIES

Capillary Alveolar Dysplasia
Pulmonary artery Sling
AVM

RESPIRATORY AIRWAYS ANOMALIES

Newman B. Pediatr Radiol 2006
The management controversy

- Prenatal diagnosis
- At birth:
  - Symptomatic cyst → surgery
    - Respiratory distress
    - Recurrent infections
    - Pneumothorax
  - Asymptomatic cyst → ?
    - Persistency ?
    - Size ?
    - Malignancy ?
- Timing for surgery → 2 weeks – 24 months
- 8% spontaneous resolution at 9 monthsa

Congenital Lung Cysts and malignancy

- Congenital cystic lung lesions: 1/8,300-35,000 births
- The large-cyst subtypes account for 60-70%
- > 90% are not neoplastic

- Childhood lung malignancies:
  - Pleuropulmonary Blastoma (PPB)
    - Most common (36% type I; 36% type II; 28% type III)
    - Rhabdomyosarcomas or progressing PPB
    - PPB Family Tumor Susceptibility Syndrome
  - Bronchioloalveolar Carcinoma (BAC)
  - Pleuropulmonary synovial sarcoma

- Estimate incidences (IPPBR data):
  - PPB: 0.35-0.65 cases per 100,000 births
  - CPAM: 2-8 cases per 100,000 births (3-22 times more common)
### Congenital Cystic Adenomatoid Malformation (CCAM)

#### Congenital Pulmonary Airway Malformation (CPAM)

- **1:25,000**
- **Bronchial obstruction?**
  - \( \rightarrow \) alveolar development
  - \( \rightarrow \) communicating cysts
- **Infection – PNX – Cancer**

<table>
<thead>
<tr>
<th>Cyst size</th>
<th>Stocker (classic)</th>
<th>Anatomic (in use)</th>
</tr>
</thead>
<tbody>
<tr>
<td>I, &gt; 2 cm</td>
<td>Macrocytic, &gt; 5 mm</td>
<td>Microcytic, &gt; 5 mm</td>
</tr>
<tr>
<td>II, &lt; 2 cm</td>
<td>Microcytic, &lt; 5 mm</td>
<td></td>
</tr>
<tr>
<td>III, solid</td>
<td>Microcytic</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Associated anomalies</th>
<th>Stocker (classic)</th>
<th>Anatomic (in use)</th>
</tr>
</thead>
<tbody>
<tr>
<td>II</td>
<td>Microcytic</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Favourable prognosis</th>
<th>Stocker (classic)</th>
<th>Anatomic (in use)</th>
</tr>
</thead>
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<tr>
<td>I</td>
<td>Macrocytic</td>
<td></td>
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</table>

<table>
<thead>
<tr>
<th>Unfavourable prognosis</th>
<th>Stocker (classic)</th>
<th>Anatomic (in use)</th>
</tr>
</thead>
<tbody>
<tr>
<td>III</td>
<td>Microcytic</td>
<td></td>
</tr>
</tbody>
</table>

## Congenital Cystic Adenomatoid Malformation (CCAM)

### Congenital Pulmonary Airway Malformation (CPAM)

<table>
<thead>
<tr>
<th>Type</th>
<th>%</th>
<th>Level of anomaly</th>
<th>X-ray</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>&lt; 5 %</td>
<td>Trachea – Mainstream Bronchi</td>
<td>Pulmonary hypoplasia – Extremely poor prognosis</td>
</tr>
<tr>
<td>1</td>
<td>60-70%</td>
<td>Mainstream Bronchi - Bronchioli</td>
<td>&gt; 2 cm cyst</td>
</tr>
<tr>
<td>2</td>
<td>15-20%</td>
<td>Bronchioli</td>
<td>Macrocysts (1-2 cm) within a microcystic mass,</td>
</tr>
<tr>
<td>3</td>
<td>5-10%</td>
<td>Bronchioli- Alveolar ducts</td>
<td>Microscopic cysts, solid aspect</td>
</tr>
<tr>
<td>4</td>
<td>&lt; 5%</td>
<td>Alveoli-Acinus</td>
<td>Macroscopic cysts + mediastinal shift</td>
</tr>
</tbody>
</table>

Updated Stocker Classification

Pulmonary Sequestration

75%

25%

60% - Thoracic aorta
20% - Abdominal aorta
15% - Multiple origins
5% - Other

Newman B. Pediatr Radiol 2006
Intralobar Pulmonary Sequestration
Extralobar Pulmonary Sequestration
Hybrid CPAM-Sequestration
Pleuropulmonary Blastoma (PPB)

- Dysembryonic malignancy (resemblance to corresponding embryonal tissue)
- >94% in children < 6 yrs
- Intrapulmonary (lung periphery) or extrapulmonary (mediastinum, diaphragm, parietal pleura)
- Stages of one malignant process on a biological continuum* from:
  - Type I: entirely cystic lesion (median age at diagnosis 9 months)
  - Type II: cystic and solid mass (mixed-pattern primitive sarcoma) (median 36 months)
  - Type III: purely solid high-grade sarcoma (median 41 months)
- Extrapulmonary localization or delayed recognition of the early cystic stage significantly worsens a child’s prognosis**

Type I PPB

- Typically presents within the first 2 yrs of life
- Large cyst/s, usually multilocular
- Peripheral lung
- Adjacent to or involving visceral pleura
- ± Respiratory distress
- ± Pneumothorax (43%)
- 10% bilateral cysts (vs 2% CPAM)
- 5% multifocal cysts
Type I PPB

• Microscopic aspects:
  – Unicystic, multilocular architecture with delicate septa
  – Small primitive mesenchymal cells within the stroma, beneath the benign epithelial lining (may regress)
  – Possible rhabdomyomatous differentiation
  – Possible small nodules of cartilage
  – Rarely anaplasia
  – No pathognomonic molecular marker of PPB
Progression of type I PPB

• **Increasing complexity from type I to type II...**
  – Progressive overgrowth and expansion of the septa by malignant cells
  – Solid nodules bulging into cysts
  – Mostly solid tumors with scattered residual cystic foci
  – 78% present anaplasia
  – 10% metastasize

• **...to type III**
  – Exclusively solid tumor
  – Multipatterned, highly malignant appearing sarcoma
  – 84% present anaplasia
  – 55% metastasize

...moreover...

• **Evidences:**
  – Recurrences of PPB I are typically advanced disease
  – “Benign lung cysts” for months/years preceding the diagnosis of advanced PPB → unrecognized type I PPB
  – 5-year Kaplan Meier Survival: 85-90% (Type I) → 60% (Type II) → 45% (Type III)

Genetic factors in PPB

- 60-75% cases are sporadic
- 25-40% cases present a genetic predisposition to dysplasia or neoplasia in the patient/family (IPPBR data)
- PPB Family Tumor Susceptibility Syndrome
PPB Family Tumor Susceptibility Syndrome

- Multifocal and/or bilateral lung cysts
- Cystic Nefroma
- Small bowel polyps
- Nodular thyroid lesions (follicular carcinoma)
- Ovarian stromal-cell tumors
- Gonadal germ-cell tumors
- Childhood sarcomas
- NF 1
The differential diagnosis of air-filled lung cyst

- “Air-filled lung cyst”: CPAM types 1, 2, and 4 + extralobar sequestration
- Neither type 2 CPAM nor extralobar sequestration are reported to be associated with malignancy
- Focus on:
  - PPB in CPAM types 1 and 4
  - BAC in CPAM type 1
# Pulmonary Cysts in Early Childhood and the Risk of Malignancy

John R. Priest, MD, 1* Gretchen M. Williams, BA, 2 D. Ashley Hill, MD, 3 Louis P. Dehner, MD, 3 and Adam Jaffé, MD, FRCP, FRCPCH, FRACP 4

## TABLE 2—Malignancies Associated With Lung Cysts in Children

<table>
<thead>
<tr>
<th>Malignancy</th>
<th>Associated cyst type</th>
<th>Number of cases &lt;20 years of age</th>
<th>Gross morphology</th>
<th>Age at diagnosis</th>
<th>Common presentations 1</th>
<th>Prognosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Type I PPB</td>
<td></td>
<td>~450</td>
<td>Cystic</td>
<td>9 mo</td>
<td>Range 0–114 mo</td>
<td>Dyspnea, pneumothorax in 40%, or incidental discovery</td>
</tr>
<tr>
<td>Type II PPB (CPAM type 4)</td>
<td></td>
<td></td>
<td>Cystic and solid</td>
<td>36 mo</td>
<td>Range 6–236 mo 3</td>
<td>Dyspnea, pneumothorax in 28%, “pneumonia,” or incidental discovery</td>
</tr>
<tr>
<td>Type III PPB</td>
<td></td>
<td></td>
<td>Solid</td>
<td>43 mo</td>
<td>Range 18–147 mo</td>
<td>Dyspnea, “pneumonia”</td>
</tr>
<tr>
<td>Bronchiolo-alveolar carcinoma (BAC) in CPAM</td>
<td></td>
<td>&lt;10</td>
<td>Cystic and solid</td>
<td>20 yr 2</td>
<td>Range 0.5–75 yr</td>
<td>Recurrent infection, hemoptyis, cough, chest pain, pleural effusion</td>
</tr>
<tr>
<td>Primary pleuropulmonary synovial sarcoma</td>
<td>Unknown</td>
<td>&lt;5</td>
<td>Cysts (blebs), cystic and solid, solid</td>
<td>30 yr</td>
<td>Range 15–72 yr</td>
<td>Dyspnea, cough, pneumothorax, chest pain, hemoptyis, hemothorax</td>
</tr>
</tbody>
</table>

1*“Pneumonia:” dyspnea, cough, fever, chest and/or abdominal pain, anorexia and/or malaise.
2 BAC not associated with CPAM has median diagnosis age 68 years.
3 Excludes an unusual patient diagnosed at 36 years. 94
4 Five-year Kaplan–Meier overall survival.
<table>
<thead>
<tr>
<th>CCAM name</th>
<th>CPAM name</th>
<th>Type 0</th>
<th>Type 1</th>
<th>Type 2</th>
<th>Type 3</th>
<th>Type 4</th>
</tr>
</thead>
<tbody>
<tr>
<td>Descriptive name</td>
<td>&quot;Acinar dyplasia or dygenesis&quot;</td>
<td>&quot;The large cyst lesion&quot;</td>
<td>&quot;The small cyst lesion&quot;</td>
<td>&quot;The adenomatoid lesion&quot;</td>
<td>&quot;The unlined cyst lesion&quot; (actually lined by flattened respiratory epithelium)</td>
<td></td>
</tr>
<tr>
<td>Postulated airway origin</td>
<td>Tracheal/bronchial</td>
<td>Bronchial/bronchiolar</td>
<td>Bronchiolar</td>
<td>Bronchiolar/alveolar duct</td>
<td>Distal acinar</td>
<td></td>
</tr>
<tr>
<td>Cystic?</td>
<td>No</td>
<td>Yes</td>
<td>Yes, multiple</td>
<td>No (or scattered)</td>
<td>Yes</td>
<td></td>
</tr>
<tr>
<td>Adenomatoid?</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>No</td>
<td></td>
</tr>
<tr>
<td>Proportion of CPAM</td>
<td>&lt;2%</td>
<td>60–70%</td>
<td>15–20%</td>
<td>5–10%</td>
<td>10%</td>
<td></td>
</tr>
<tr>
<td>Unique features</td>
<td>All lobes involved; incompatible with life</td>
<td>More common in males</td>
<td>Tension pneumothorax</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Typical age at presentation</td>
<td>Birth</td>
<td>In utero, if large, to many years of age, if small</td>
<td>1st month of life</td>
<td>In utero; or at birth</td>
<td>Newborn to 6 years, or rarely much later</td>
<td></td>
</tr>
<tr>
<td>Presentation</td>
<td>Lungs will not aerate</td>
<td>Non-pulmonary distress, mediastinal shift, or later. incidentally, or cough + fever + infection, or emergence of tumor</td>
<td>Non-pulmonary anomalies may supersede lung abnormalities</td>
<td>May be stillborn, or severe neonatal respiratory distress</td>
<td>Respiratory distress ± tension pneumothorax, or infection/pneumonia, or incidental finding, or emergence of tumor</td>
<td></td>
</tr>
<tr>
<td>Lobar involvement</td>
<td>All lobes involved</td>
<td>One lobe in 95%; rarely bilateral</td>
<td>Usually one lobe</td>
<td>Entire lobe or lung</td>
<td>Usually one lobe</td>
<td></td>
</tr>
<tr>
<td>Associated anomalies</td>
<td>CV anomalies, renal hypoplasia, focal dermal hypoplasia</td>
<td>None</td>
<td>CV anomalies, diaphragmatic hernia, extralobar sequestration, renal agenesis or dysgenesis</td>
<td>N/A</td>
<td>Patient or familial childhood neoplasia and dysplasia suggests PPB (see Table 3)</td>
<td></td>
</tr>
<tr>
<td>Lesion and cyst size</td>
<td>Lungs small</td>
<td>1–10 cm</td>
<td>0.5–2.0 cm</td>
<td>Entire lobe or lung</td>
<td>Large multicellular cysts</td>
<td></td>
</tr>
<tr>
<td>Malignancy risk</td>
<td>No</td>
<td>BAC</td>
<td>No</td>
<td>No</td>
<td>PPB</td>
<td></td>
</tr>
</tbody>
</table>

1Sources: Stocker et al.,42 Stocker,41 Bush,1 Langston,39 and International PPB Registry unpublished data.
Differential diagnosis

- 66% of PPB are associated with lung cysts (IPPBR data).
- CPAM type 4 and cystic PPB are not easily distinguished from each other and have many overlapping clinical (PNX), radiographic, and pathological features.
- Hypothesis: cysts associated with PPB are not CPAM but may be separate diagnosable pathologic entities (Type I “cystic” PPB).
- Complete resection is probably the best option for both these/this entities/y.

• PPB is not a pre-existing CPAM which has undergone “malignant transformation”.

• Cystic PPB is the earliest manifestation of a sequence in which low-grade cystic PPB evolves over 2-4 years to a high-grade, solid sarcomatous disease.

Surgery is strongly advised whenever any PPB-associated condition is present.

**Risk factors for PPB**

<table>
<thead>
<tr>
<th>Risk factor for PPB</th>
<th>Degree of PPB risk</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Pulmonary manifestations</strong></td>
<td></td>
</tr>
<tr>
<td>Pneumothorax presentation</td>
<td>High</td>
</tr>
<tr>
<td>Bilateral lung cysts</td>
<td>High</td>
</tr>
<tr>
<td>Multifocal lung cysts</td>
<td>High</td>
</tr>
<tr>
<td>Familial lung cysts</td>
<td>High</td>
</tr>
<tr>
<td><strong>Associated conditions (patient or family)</strong></td>
<td></td>
</tr>
<tr>
<td>Renal cystic disease, especially cystic nephroma</td>
<td>High</td>
</tr>
<tr>
<td>Small bowel polyps</td>
<td>High</td>
</tr>
<tr>
<td>Additional childhood cancer or dysplasia, especially mesenchymal proliferations</td>
<td>High</td>
</tr>
<tr>
<td>Gonadal tumors</td>
<td>High</td>
</tr>
<tr>
<td>Any childhood cancer</td>
<td>High</td>
</tr>
<tr>
<td>Nodular thyroid hyperplasia or cancer</td>
<td>Unknown</td>
</tr>
</tbody>
</table>
Management

Prenatal/Neonatal “lung cyst”

Symptoms

No symptoms

SURGERY

Positive family history

Negative family history

Chest CT
Renal US

Multifocal/bilateral cysts
PNX
Renal mass

Single lung cyst, no PNX,
normal kidneys: repeat
investigations in 3/12

Multifocal/bilateral cysts
PNX
Renal mass

Unchanged

Careful education
Counseling
Ongoing family assessment

Follow up

• Pathological examination with no evidence of PPB or PPB-like changes $\Rightarrow$ no need for continued surveillance

• In case pathological examination reveals PPB $\Rightarrow$ management and proper strategy designed by an oncology team
BronchioloAlveolar Carcinoma (BAC)

- Adenocarcinoma of the lung
- 3-4% of all adult non-small-cell lung cancers
- Median age at diagnosis: 68 yrs
- The majority of BAC are not associated with congenital lung cysts
- Association with CPAM type I (median age at diagnosis: 18 yrs)
# BronchioloAlveolar Carcinoma (BAC)

<table>
<thead>
<tr>
<th>Reference</th>
<th>Age</th>
<th>Sex</th>
<th>Symptoms</th>
<th>Location</th>
<th>Treatment</th>
<th>Follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>Prichard et al.</td>
<td>30</td>
<td>F</td>
<td>Productive cough</td>
<td>LLL</td>
<td>Lobectomy</td>
<td>WI</td>
</tr>
<tr>
<td>Hurley et al.</td>
<td>33</td>
<td>F</td>
<td>Productive cough, dyspnea, hemoptysis</td>
<td>RML</td>
<td>Right middle and upper lobectomy</td>
<td>DF after 9 mo</td>
</tr>
<tr>
<td>Sheffield et al.</td>
<td>18</td>
<td>M</td>
<td>Hemoptysis, dyspnea</td>
<td>LUL</td>
<td>Lobectomy</td>
<td>WI</td>
</tr>
<tr>
<td>Benjamin and Cahill</td>
<td>19</td>
<td>M</td>
<td>Productive cough</td>
<td>LLL</td>
<td>Pneumonectomy</td>
<td>Died 4 y later</td>
</tr>
<tr>
<td>Morresi et al.</td>
<td>20</td>
<td>F</td>
<td>Incidental finding during routine examination</td>
<td>LUL</td>
<td>Partial lobectomy</td>
<td>DF after 8 y</td>
</tr>
<tr>
<td>Ribeiro et al.</td>
<td>41</td>
<td>F</td>
<td>Dyspnea</td>
<td>LLL</td>
<td>Lobectomy</td>
<td>DF after 8 y</td>
</tr>
<tr>
<td>Ribeiro et al.</td>
<td>42</td>
<td>F</td>
<td>Incidental finding during routine examination</td>
<td>LLL</td>
<td>Lobectomy</td>
<td>DF after 2 y</td>
</tr>
<tr>
<td>Kaslovska et al.</td>
<td>11</td>
<td>M</td>
<td>Chest pain and multiple nodules throughout both lungs</td>
<td>RUL</td>
<td>Lobectomy</td>
<td>WI</td>
</tr>
<tr>
<td>Olive et al.</td>
<td>6</td>
<td>M</td>
<td>Paroxysmal cough, intermittent hemoptysis, and vague chest pain</td>
<td>LLL</td>
<td>Lobectomy</td>
<td>RML, metastasis with resection, DF after 7 y</td>
</tr>
<tr>
<td>Granata et al.</td>
<td>11</td>
<td>M</td>
<td>Recurrent pneumonia</td>
<td>LLL</td>
<td>Lobectomy</td>
<td>DF after 15 y</td>
</tr>
<tr>
<td>Sufi et al.</td>
<td>17</td>
<td>M</td>
<td>Dyspnea on exertion</td>
<td>LLL</td>
<td>Lobectomy</td>
<td>DF after 3 mo</td>
</tr>
<tr>
<td>MacSweeney et al.</td>
<td>6</td>
<td>M</td>
<td>Wheezing</td>
<td>RUL</td>
<td>WI</td>
<td>DF after 16 y</td>
</tr>
<tr>
<td>13</td>
<td>M</td>
<td>WI</td>
<td></td>
<td>LLL</td>
<td>WI</td>
<td>DF after 11 y</td>
</tr>
<tr>
<td>18</td>
<td>M</td>
<td>WI</td>
<td></td>
<td>LLL</td>
<td>WI</td>
<td>DF at 2003</td>
</tr>
<tr>
<td>30</td>
<td>M</td>
<td>WI</td>
<td></td>
<td>RLL</td>
<td>WI</td>
<td>DF after 4 y</td>
</tr>
<tr>
<td>36</td>
<td>M</td>
<td>WI</td>
<td></td>
<td>NK</td>
<td>WI</td>
<td>NK</td>
</tr>
<tr>
<td>Ioachimescu et al.</td>
<td>6</td>
<td>M</td>
<td>Pleuritic chest pain, cough, intermittent hemoptysis, weight loss, pulmonary nodules</td>
<td>LLL</td>
<td>Lobectomy</td>
<td>BAC terminating in invasive AC with metastasis after 15 y</td>
</tr>
<tr>
<td>12</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Lantuchov et al.</td>
<td>60</td>
<td>M</td>
<td>Hemoptysis</td>
<td>RLL</td>
<td>Lobectomy</td>
<td>Mixed AC with BAC pattern</td>
</tr>
<tr>
<td>13</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>32</td>
<td>M</td>
<td></td>
<td>Recurrent infections, hemoptysis</td>
<td>RLL</td>
<td>Biopsy of the right lung</td>
<td>Multifocal mucinous BAC</td>
</tr>
<tr>
<td>This case</td>
<td>8</td>
<td>F</td>
<td>Recurrent pneumonia</td>
<td>LLL</td>
<td>Lobectomy</td>
<td>Metastatic disease after 2 y</td>
</tr>
</tbody>
</table>

F indicates female; M, male; DF, disease free; LUL, left upper lobe; LLL, left lower lobe; RUL, right upper lobe; RML, right middle lobe; RLL, right lower lobe; NK, not known; WI, without information.
Primary Pleuropulmonary Synovial Sarcoma

- Rare cystic intrathoracic sarcoma
- Adolescents and older patients
- Solid or cystic proliferation
- ± pneumothorax
- t(X;18) SYT-SSX gene fusion
Surgical approach
Congenital pulmonary malformations: IGG experience

1991-2009: 89 included

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>n° (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pulmonary Seq</td>
<td>29 (33)</td>
</tr>
<tr>
<td>Intralobar</td>
<td>23 (26)</td>
</tr>
<tr>
<td>Extralobar</td>
<td>6 (7)</td>
</tr>
<tr>
<td>CPAM</td>
<td>22 (25)</td>
</tr>
<tr>
<td>Macrocystic</td>
<td>12 (14)</td>
</tr>
<tr>
<td>Microcystic</td>
<td>10 (11)</td>
</tr>
<tr>
<td>Hybrid SEQ/CPAM</td>
<td>1 (1)</td>
</tr>
<tr>
<td>Bronchogenic cyst</td>
<td>16 (18)</td>
</tr>
<tr>
<td>CLE</td>
<td>11 (12)</td>
</tr>
<tr>
<td>Other cysts</td>
<td>10 (11)</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td>89 (100)</td>
</tr>
</tbody>
</table>
Congenital pulmonary malformations: IGG experience

- Pulmonary Seq
- CPAM
- Bronchogenic cyst
- CLE
- Other cysts

- Wedge resections
- Lobectomies
- Total
Complications

• No intraoperative complications

• Postoperative complications:
  – Bleeding 3 (3.4 %)
  – Disease persistency / recurrency 1 (1.1 %)
  – PNX 1 (1.1 %)

• Cancer: 2 (2.3%)
  – 1 Cystic PPB excised. Relapsed to high grade PPB → exitus
  – 1 BAC excised, still alive, disease-free
Follow up

Spirometry

Quality of life

$r = 0.34, p = 2$

Apolone G, et al. Questionario sullo stato di salute SF-12
Conclusions & Recommendations

• Focus on pathogenesis and cancer development
• Early diagnosis → early treatment
  – Surgery can be delayed until the child is 9/12 to allow for possible resolution without risking the emergence of malignancy
• Minimal access surgery
  – Thoracoscopy vs Thoracothomy
• Sparing surgery
  – Segmentectomy vs Lobectomy
• Low complication rate
• Quality of life

Thank you!