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# **The rare case of bilateral polycystic kidney disease associated with congenital cystic adenomatoid malformation.**

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# Congenital Cystic Adenomatoid Malformation (CCAM)



- rare congenital malformation
- anomalous fetal development of terminal respiratory structures.
- dysplastic, multicystic masses interfere with alveolar development.

*(Ann Thorac Surg 2007;83:e2)*

# CCAM

- 25% of all bronchopulmonary congenital malformation
- 1/25.000-35.000 pregnancies
- No gender predominance
- No connection with mother's age
- No connection with inborn infection
- Possibility of malignant transformation

1. J. Pediatr. Surg., 2010, 45 (2), e25-8
2. Fetal Diagn. Ther., 2001, 16 (3), 178-186
3. Ped. Pol., 2002, 77 (8), 689-700
4. J. Pediatr. Surg., 2010, 45 (6), 1086-1089

# CCAM - Stocker classification

I	<ul style="list-style-type: none"><li>■ Single or multiple air filled cysts, often more than 2 cm.</li><li>■ Respiratory epithelium</li></ul>
II	<ul style="list-style-type: none"><li>■ Cysts smaller than 2 cm and mixed with solid tissue.</li><li>■ Ciliated cuboidal or columnar epithelium</li></ul>
III	<ul style="list-style-type: none"><li>■ A solitary mass</li><li>■ Bronchiole-like structures -ciliated cuboidal epithelium</li><li>■ Masses of alveolus-sized structures - nonciliated cuboidal epithelium</li></ul>

# Coexistence with other diseases 4-26%\*

- Anasarca and polyhydramnios in fetus
- bronchopulmonary sequestration
- Hydrocephalus
- Diaphragmatic hernia
- intestinal atresia
- Tracheo-esophgal fistula
- Tetralogy of Fallot
- polycystic kidney disease

# Literature

## Only 4 cases of CCAM in coexistence with PKD

- 1971 Roloff and others

Bilateral CCAM and cysts in renal medulla in 20-month girl

- 1997 Jamet F and others

CCAM and renal cysts in fetus

- 1985 Atamanov

Cysts of lungs, kidneys and thyroid

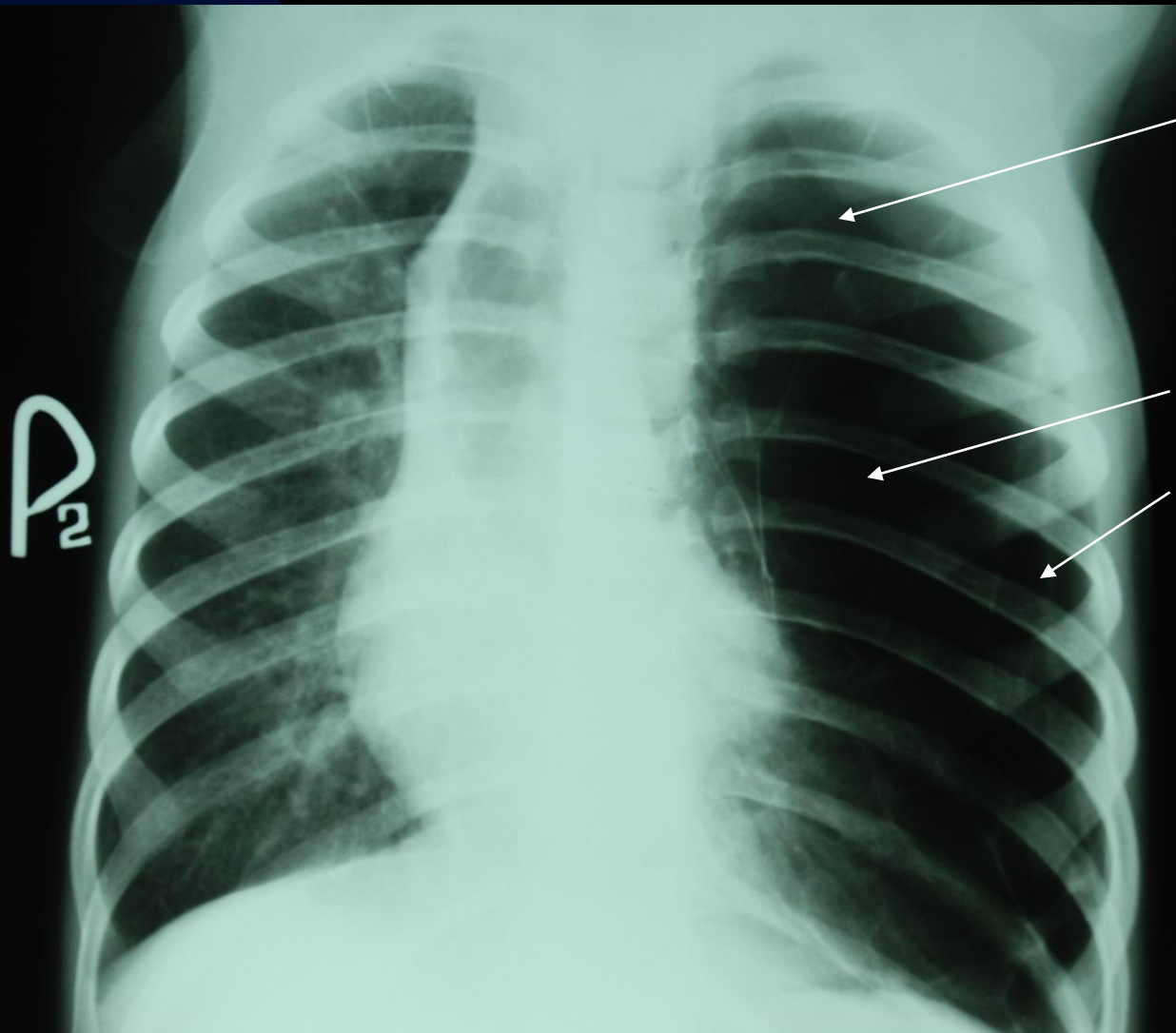
- 1987 Graham and others

„hamartomata of lungs and kidney”

# Case report

- A 2-year old girl
- Admitted due to left-side pneumothorax and pneumonia
  
- Born in 40 hbd
  - Apgar 10 points
  - No respiratory distress

# X-ray



Distension in the  
upper part of the  
lung

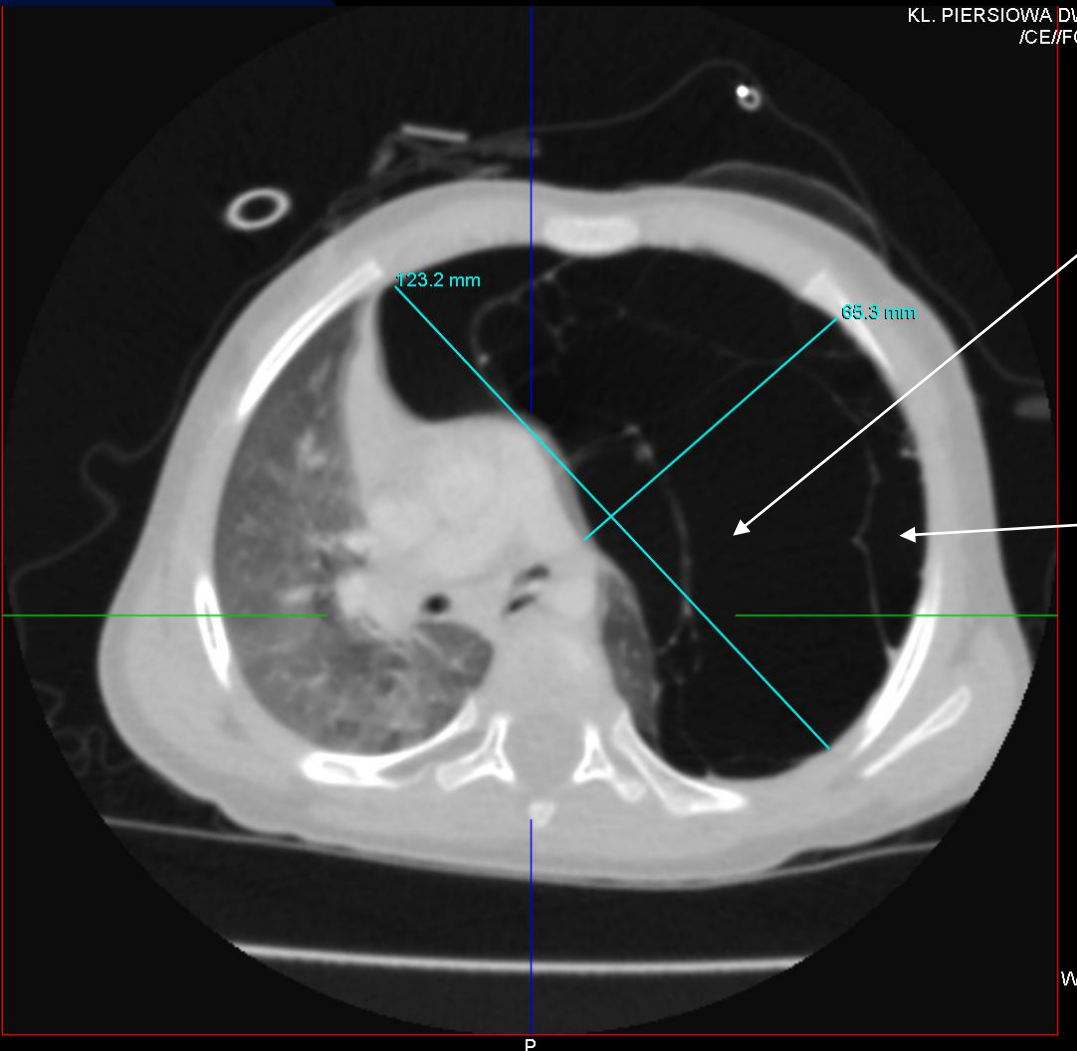
Blurry vessel image

Pleura line

Pneumonia image  
bilaterally



# CT scan of the patient



Air bulbs (up to 41mm $\emptyset$ ), fine septum between

Air in the pleural space

Mediastinal shift to the right

# Toracotomy

- Histopathology Nr 270-273/09
- One big cyst and multiple smaller cystic structures
- Columnar epithelium
- Cuboidal epithelium

# Kidney observation in USG

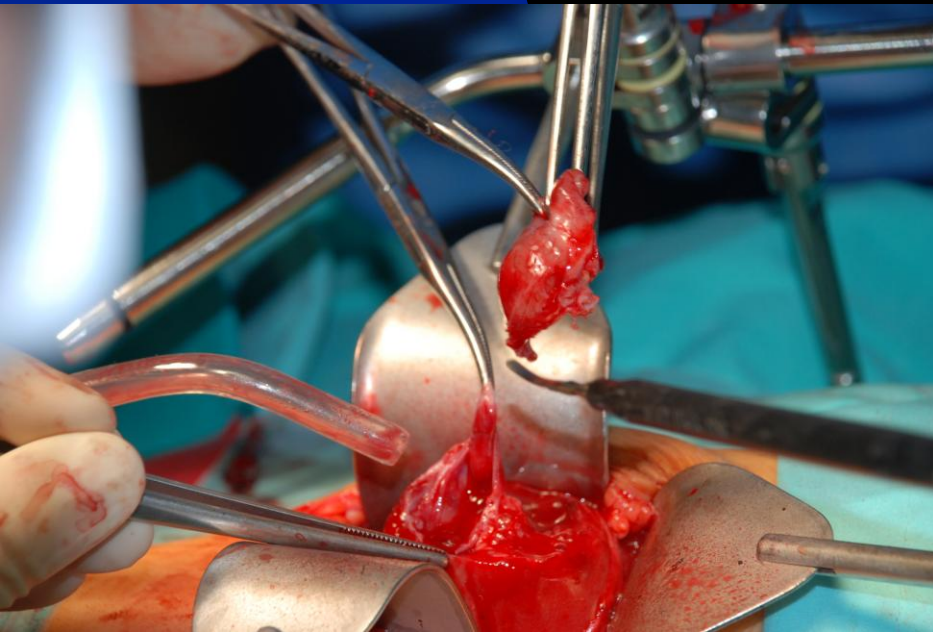
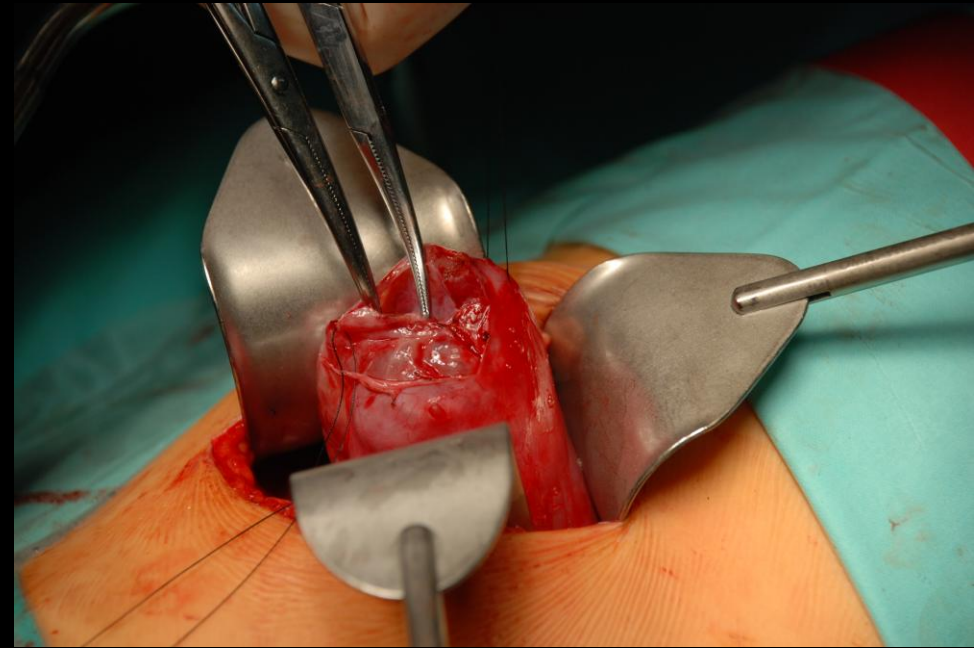
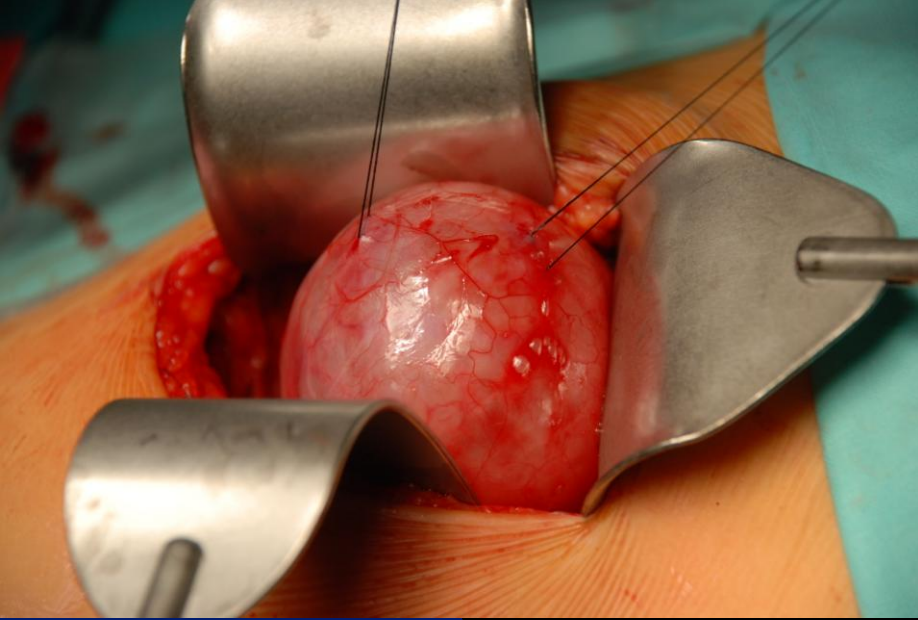


**Further examinations and  
USG controls of the left kidney**



**Increasing of cystc**

# Excision of lower pole of left kidney



Multicystic cyst of the left kidney – intraoperative photograph

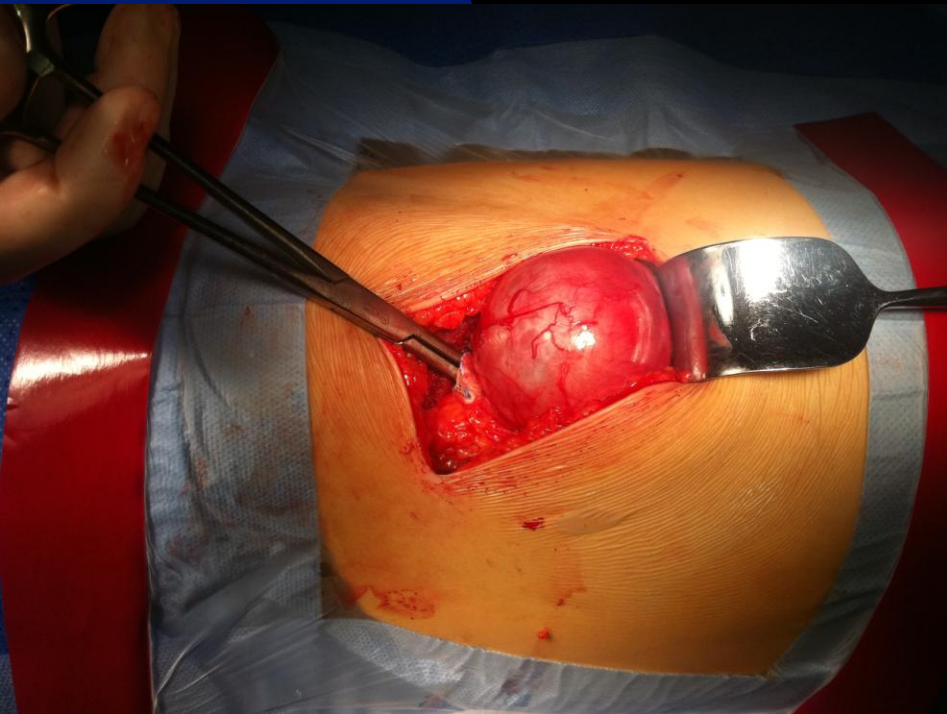
**9-month observation of right kidney**

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graph TD; A[9-month observation of right kidney] --> B[➤ Increasing of malformations<br/>➤ Pain symptoms]; B --> C[Surgical intervention];
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- **Increasing of malformations**
- **Pain symptoms**

**Surgical intervention**

# Resection of lower pole of the right kidney



Postoperative material

Nr 1715-19/10

- Multiple cysts
- Cuboidal epithelium

# USG control



# Conclusions

- Presence of CCAM should be an indication to vast diagnostic procedure in search of other inborn abnormalities
- Excision of the lung's lesion is considered a standard procedure of choice
- One single renal cyst in children with CCAM may give a suspicion of polycystic kidney disease and require further controls.



Thank You for  
Your attention

