



# An infant with fever and tachypnoea: diagnosis at first sight?

Edith M. van den Beukel - Bakker<sup>1,2</sup> and Marielle W.H. Pijnenburg<sup>2</sup>

<sup>1</sup>Dept of Paediatrics, Maastricht ziekenhuis, Rotterdam, The Netherlands. <sup>2</sup>Erasmus MC-Sophia Children's Hospital, University Medical Center Rotterdam, Dept of Paediatrics/Paediatric Respiratory Medicine and Allergology, Rotterdam, The Netherlands.

Corresponding author: Edith M. van den Beukel - Bakker (marijevandenbeukel@gmail.com)



Shareable abstract (@ERSpublications)

**Complicated pneumonia can resemble an infection in congenital pulmonary abnormalities such as CPAM or sequestration. Both have an increased risk of a complicated course. Imaging can support treatment decisions and may avoid unnecessary surgical intervention.** <https://bit.ly/3IMU9aL>

**Cite this article as:** van den Beukel - Bakker EM, Pijnenburg MWH. An infant with fever and tachypnoea: diagnosis at first sight? *Breathe* 2022; 18: 220002 [DOI: 10.1183/20734735.0002-2022].

Copyright ©ERS 2022

*Breathe* articles are open access and distributed under the terms of the Creative Commons Attribution Non-Commercial Licence 4.0.

Received: 7 Jan 2022  
Accepted: 22 March 2022

A 6-month-old girl presented at a local hospital with a fever of 39 °C, agitation, and general malaise. The fever started more than a month before presentation when she had chickenpox (varicella). However, even after all skin lesions resolved, fever persisted.

## Task 1

Which complications can occur following a varicella zoster infection?

[Go to Answers >>](#)

On physical examination, the girl appeared uncomfortable and was mildly tachypnoeic with a respiratory rate of 42 breaths per min. She had a blood pressure of 90/60 mmHg, heart rate of 155 beats per min, peripheral oxygen saturation ( $S_{pO_2}$ ) of 96% in room air and temperature of 39.1 °C.

Except for decreased breath sounds on the right side compared to the left, lung auscultation was normal. There were no abnormalities on examination of the heart and abdomen. The neurological examination was also normal.

Laboratory investigations revealed mild anaemia (haemoglobin 5.3 mmol·L<sup>-1</sup>), elevated leukocytes (28×10<sup>9</sup> per L), slightly raised platelet counts (461×10<sup>9</sup> per L) and an elevated C-reactive protein (CRP) of 159 mg·L<sup>-1</sup>.

Because of the persisting fever and tachypnoea, a chest radiograph was performed (figure 1).

## Task 2

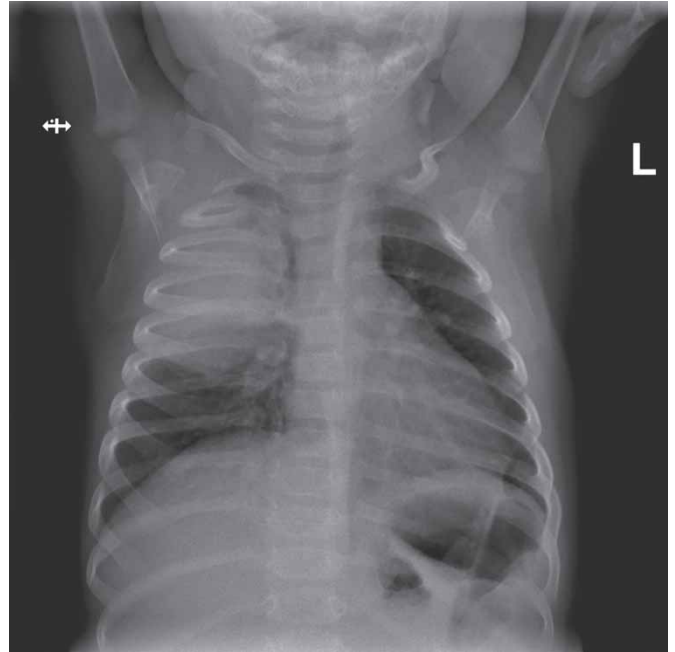
Please describe any abnormalities seen on the chest radiograph (figure 1).

[Go to Answers >>](#)

Because of the fever, elevated CRP and abnormalities on the chest radiograph, a diagnosis of lobar pneumonia was suspected. Treatment with oral amoxicillin was started and the patient was discharged home. The mild anaemia was considered to be related to the infection, possibly combined with physiologically lower values in infants at the age of 6 months. A conservative approach was advised and the parents were informed that laboratory tests should be repeated once the girl had recovered.

However, after 4 days the patient presented again at the emergency department because there had been no clinical improvement since the start of the antibiotics. She still had a fever up to 39 °C and seemed unwell.





**FIGURE 1** Chest radiograph at first presentation.

Physical examination was unchanged, with persisting tachypnoea (44 breaths per min) and decreased breath sounds over the right lung. There were mild retractions and no other signs of dyspnoea.

Laboratory investigations showed an improvement in CRP ( $81 \text{ mg}\cdot\text{L}^{-1}$ ), however the anaemia and leukocytosis remained unchanged. A second chest radiograph was performed (figure 2). This showed an increase of the consolidation, with few air bronchograms. There was a shift of mediastinal structures to the left. The major fissure on the right was displaced caudally.



**FIGURE 2** Second chest radiograph, performed when the patient returned to the hospital because of persistence of symptoms.

**Task 3**

What is your differential diagnosis?

[Go to Answers >>](#)

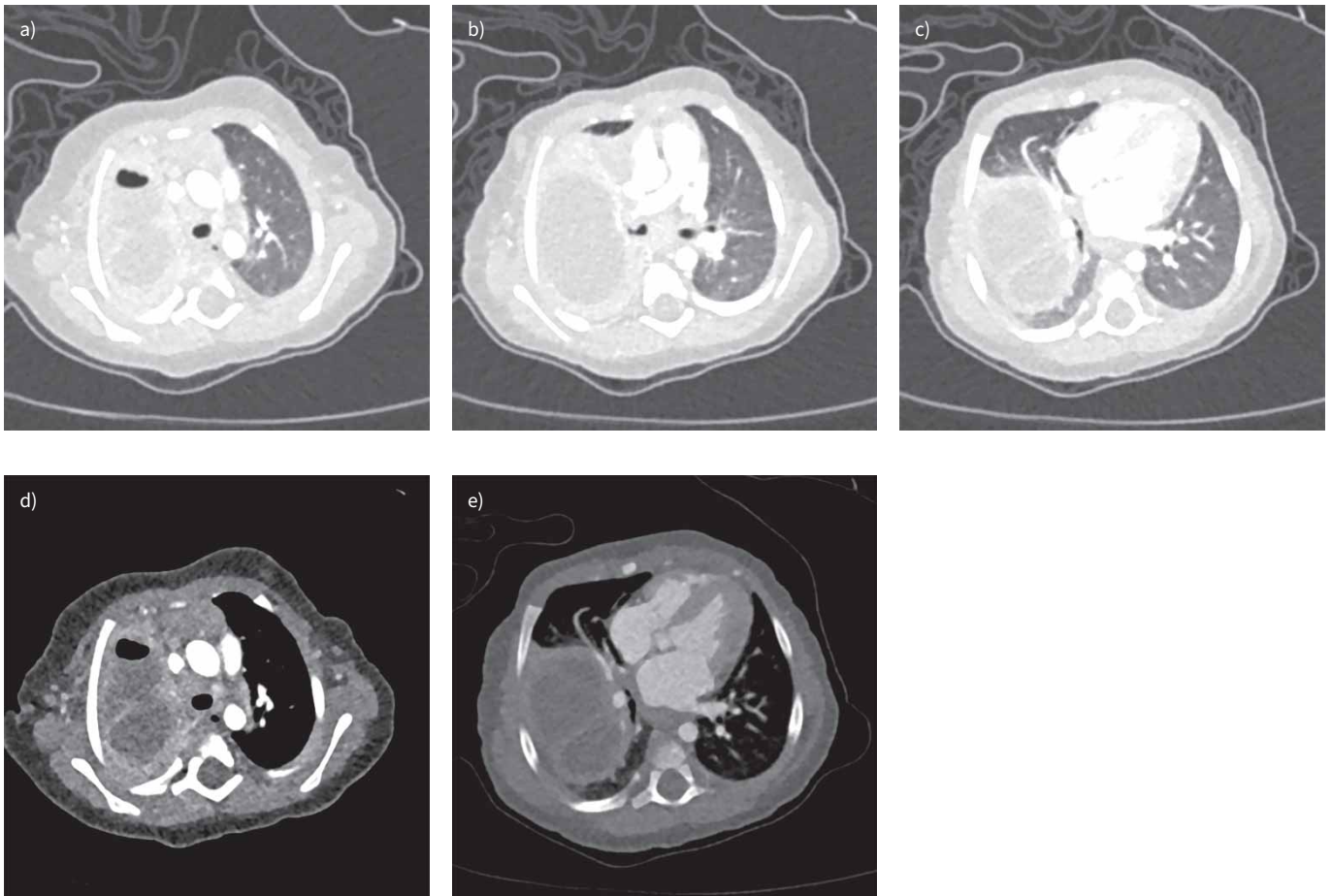
Since there was persistent fever and an increase of abnormalities on the chest radiograph despite treatment with oral antibiotics, the patient was admitted for intravenous cefuroxime. Prior to the start of the antibiotics a blood culture was taken. However, this culture remained negative. Attempts were made to obtain a sputum culture; however, due to the young age of the patient this did not succeed. A chest ultrasound was performed which revealed a large intrapulmonary mass with debris inside and a visible wall. The radiologist concluded that the abnormality could be a lung abscess or an infection in a congenital anatomical abnormality. A malignant tumour could not be ruled out, although this seemed less likely because of the appearance of the abnormalities on ultrasound and the signs and symptoms of the patient, which fitted an infectious cause of the problems.

**Task 4**

What would be your next step?

[Go to Answers >>](#)

The next day, the girl was transferred to our tertiary care hospital and a chest CT scan was performed (figure 3a). The CT scan was performed after the girl was breastfed and fell asleep. Using an inflatable mattress to minimise movement, no sedation or anaesthesia was needed.



**FIGURE 3** Three slides of the lung window of the CT scan of the chest (top row): a) just below the aortic arch, b) at the level of the pulmonary artery, just below the carina, and c) at the level of the right middle and lower lobe. d, e) Two slides of the mediastinal window on the CT scan, which show the fluid-filled abnormalities: possible cystic lesions of an infected CPAM.

The CT scan showed a large round intrapulmonary abnormality, located mostly in the right upper lobe. There were fluid and air collections inside the abnormality with partial compression of the right main bronchus and the lobar bronchi to the middle and lower lobes. The left lung appeared normal. Our differential diagnosis was an infected CPAM or a pneumonia with lung abscesses (figure 3b).

The paediatric chest surgeon was consulted and agreed with our differential diagnosis.

Sputum cultures were obtained; however, these remained negative. It should be taken into consideration that at this point in time the patient had already been treated with oral antibiotics for 4 days and intravenously for 1 day.

Since CPAM was considered, we also asked whether prenatal ultrasound was performed. The parents informed us that ultrasounds were performed at 12 weeks, 20 weeks and 31 weeks of gestation, and no abnormalities were seen. Although most CPAMs are detected prenatally in developed countries, a normal ultrasound during pregnancy does not exclude a possible CPAM. Post-natal diagnosis is mainly made after pneumonia in patients with no antenatal diagnosis [1]. A retrospective study of prenatal and paediatric data for cases of suspected CPAM showed a sensitivity of prenatal detection of 81% [2].

### Task 5

How would you treat this patient?

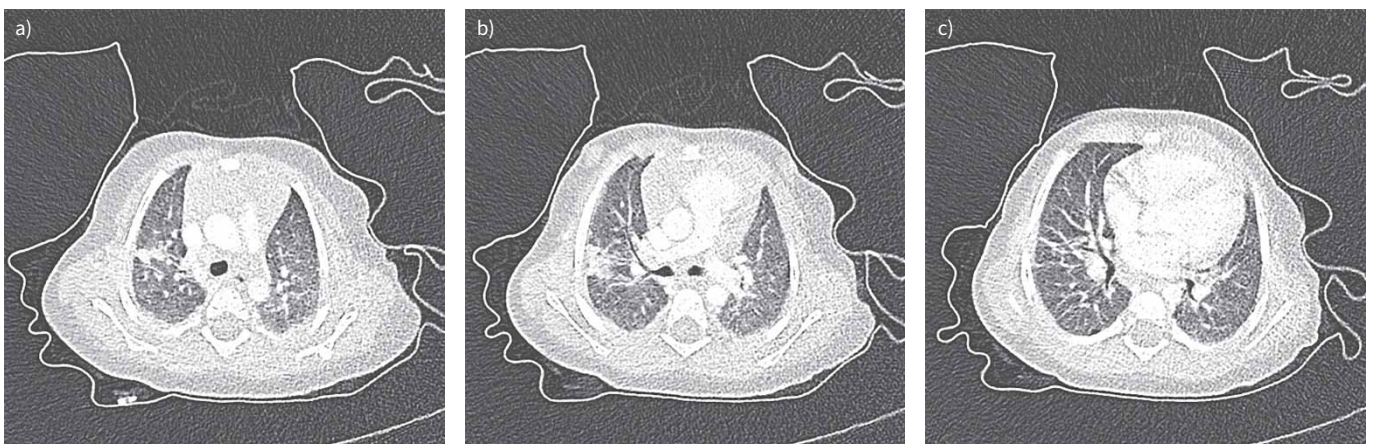
[Go to Answers >>](#)

The patient was admitted and treated with intravenous cefuroxime. Within 1 week of treatment, the patient was free of fever. Laboratory tests were repeated and showed a decrease in CRP to  $45 \text{ mg} \cdot \text{L}^{-1}$  and normalisation of the leukocyte count. She was treated with intravenous antibiotics for a total of 2 weeks and was discharged home with oral antibiotics for another 4 weeks. Parents were informed of the likely diagnosis of CPAM and resection was discussed after full recovery from the infection.

We performed a new CT scan 2 months later as part of the pre-operative assessment before lobectomy (figure 4). To our surprise, the CT scan showed a remarkable improvement and only some post-infectious fibrotic changes were seen. There were no signs of CPAM or other anatomical abnormalities. During 18 months of follow-up the child is doing well and has had no pulmonary infections or other respiratory problems. The mild anaemia resolved spontaneously, as was expected.

### Discussion

Community-acquired pneumonia (CAP) is an important cause of morbidity and mortality in children. Worldwide, ~750 000 children younger than 5 years of age die of pneumonia annually (14% of all deaths



**FIGURE 4** Second CT scan of the chest, performed 2 months later and after prolonged antibiotic treatment. Three slides are shown at similar positions to those in figure 3: a) just below the aortic arch, b) at the level of the pulmonary artery, just below the carina, and c) at the level of the right middle and lower lobe.

aged <5 years), with the highest death rates in South Asia and sub-Saharan Africa [3]. Risk factors for CAP are younger age, history of premature birth, immunodeficiency, chronic respiratory disease and neurological disability. Exposure to tobacco smoke and other indoor and outdoor pollutants also increases the risk of CAP [4].

Complications of pneumonia include pleural empyema, lung abscess and necrotising pneumonia. Congenital pulmonary abnormalities, such as CPAM, sequestration or bronchogenic cysts, may present with infection and have an increased risk of a complicated course of pneumonia [5].

### **CPAM**

In our patient, CPAM was suspected because of the cystic abnormalities on the CT scan and the complicated course of the airways infection.

CPAM, previously known as congenital cystic adenomatoid malformation (CCAM), is a rare developmental malformation of the lower respiratory tract. The incidence of CPAMs is estimated to be 1 per 25 000–35 000 live births [6, 7]. However, with the advent of prenatal ultrasound, it is likely that the true incidence may have been underestimated because previously undiagnosed lesions are now being detected before birth [8].

CPAMs result from abnormalities of branching morphogenesis of the lung. The different types of CPAMs are thought to originate at different levels of the tracheobronchial tree and at different stages of lung development, possibly influenced by *in utero* airway obstruction and/or atresia [9]. The molecular mechanisms resulting in CPAM formation are unknown, but may include an imbalance between cell proliferation and apoptosis during organogenesis [10, 11].

There is consensus that symptomatic CPAMs should be resected, whereas the management of asymptomatic CPAMs is still controversial and subject to discussion [12]. In asymptomatic CPAMs, some prefer to resect the lesion before any symptoms occur, because surgery after symptoms have occurred has more risks of complications. Furthermore, the potential for compensatory lung growth is thought to be optimal in the first 2 years of life [13, 14]. Also, questions exist about the long-term risk of malignancy in unresected CPAM lesions, such as the small associated risk of developing pleuropulmonary blastoma [15].

Others adopt a conservative strategy, recognising the surgical risks and the potential of over-treatment [1]. Additionally, a retrospective study showed that age at the time of lobectomy did not influence the spirometry results at a mean age of 10 years [16, 17].

Although we expected a CPAM in our patient, it eventually turned out to be a complicated pneumonia with formation of multiple abscesses and/or cavitations.

### **Pulmonary abscess**

One of the complications of pneumonia can be a pulmonary abscess. This is an accumulation of inflammatory cells, with tissue destruction or necrosis, leading to one or more cavities. Predisposing factors are inadequate or delayed treatment of lobar pneumonia, pulmonary aspiration, airway obstruction or a congenitally abnormal lung. Clinical manifestations are nonspecific and include fever, cough, dyspnoea, chest pain, anorexia, haemoptysis, and putrid breath.

Treatment of a pulmonary abscess consists of a prolonged course of antibiotics. In complicated pneumonia, antibiotics should be started intravenously. However, it is not exactly known when patients can be transferred from intravenous to oral antibiotic therapy. The British Thoracic Society guidelines advise that oral treatment should be considered if there is clear evidence of improvement [18]. In 80–90% of patients, treatment with antibiotics is successful. However, some patients may need an intervention, such as needle aspiration, percutaneous catheter drainage (interventional radiology), or even lobectomy [3, 19].

### **Conclusion**

In conclusion, the complicated pneumonia in our patient mimicked an infected CPAM. Our patient recovered from this complicated pneumonia with prolonged treatment with (intravenous) antibiotics. In children presenting with a complicated pneumonia, we suggest a conservative approach with antibiotics and supportive treatment. Video-assisted thoracic surgery and other surgical procedures should be reserved for carefully selected patients only.

**Answer 1**

Complications of varicella zoster can include:

- Varicella zoster pneumonia
- Bacterial pneumonia (*Staphylococcus aureus* and *Streptococcus*)
- Bacterial infections of the skin and soft tissues in children, including invasive Group A streptococcal infections
- Encephalitis, cerebellar ataxia

<< Go to Task 1

**Answer 2**

The chest radiograph shows a large consolidation in the right upper lobe. Few air bronchograms are seen. The left lung appears normal. No pleural fluid is visualised, and no pneumothorax. The heart is of normal size and position. There are no abnormalities of the bones of the thorax, spine and upper arms.

<< Go to Task 2

**Answer 3**

At this point in time, our differential diagnosis included:

- Lobar pneumonia, caused by a microorganism that is not susceptible to amoxicillin
- Pneumonia with development of an abscess
- Necrotising pneumonia
- Infection in a pre-existing anatomical abnormality, such as congenital pulmonary airway malformation (CPAM) or bronchogenic cyst
- Malignancy
- Immune deficiency and infection with an opportunistic organism
- Aspiration pneumonia

<< Go to Task 3

**Answer 4**

Since an abscess or an infection in an anatomical abnormality was suspected, the patient was transferred to a tertiary care centre. Additional radiological investigations were considered, and a computed tomography (CT) scan of the chest was performed. Also, consultation with the paediatric surgeons was initiated as the abscess might need surgical drainage.

<< Go to Task 4

**Answer 5**

An infection in an anatomical abnormality in the lung should be treated with intravenous antibiotics before any surgery or intervention can be considered. Any operation in an infected area has a higher risk of complications such as bleeding, persistent bronchopleural fistula or persistent pneumothorax.

<< Go to Task 5

Acknowledgements: Radiology images provided by Imaging and trial bureau Erasmus MC, Rotterdam, The Netherlands.

Conflict of interest: E.M. van den Beukel - Bakker is a member of a committee that updates/rewrites the Dutch guideline on bronchiolitis (supported by NVK: Nederlandse Vereniging voor Kindergeneeskunde), from whom they have received an expense allowance for three meetings with the committee. M.W.H. Pijnenburg has nothing to disclose.

**References**

- 1 Leblanc C, Baron M, Desselas E, *et al.* Congenital pulmonary airway malformations: state-of-the-art review for pediatrician's use. *Eur J Pediatr* 2017; 176: 1559–1571.
- 2 Gornall AS, Budd JL, Draper ES, *et al.* Congenital cystic adenomatoid malformation: accuracy of prenatal diagnosis, prevalence and outcome in a general population. *Prenat Diagn* 2003; 23: 997–1002.
- 3 World Health Organization. Pneumonia. Date last updated: 11 November 2021. [www.who.int/news-room/fact-sheets/detail/pneumonia](http://www.who.int/news-room/fact-sheets/detail/pneumonia)
- 4 Dherani M, Pope D, Mascarenhas M, *et al.* Indoor air pollution from unprocessed solid fuel use and pneumonia risk in children aged under five years: a systematic review and meta-analysis. *Bull World Health Organ* 2008; 86: 390C–398C.

- 5 de Benedictis FM, Kerem E, Chang AB, *et al.* Complicated pneumonia in children. *Lancet* 2020; 396: 786–798.
- 6 David M, Lamas-Pinheiro R, Henriques-Coelho T. Prenatal and postnatal management of congenital pulmonary airway malformation. *Neonatology* 2016; 110: 101–115.
- 7 Laberge JM, Puligandla P, Flageole H. Asymptomatic congenital lung malformations. *Semin Pediatr Surg* 2005; 14: 16–33.
- 8 Cass DL, Olutoye OO, Cassady CI, *et al.* Prenatal diagnosis and outcome of fetal lung masses. *J Pediatr Surg* 2011; 46: 292–298.
- 9 Riedlinger WF, Vargas SO, Jennings RW, *et al.* Bronchial atresia is common to extralobar sequestration, intralobar sequestration, congenital cystic adenomatoid malformation, and lobar emphysema. *Pediatr Dev Pathol* 2006; 9: 361–373.
- 10 Cass DL, Quinn TM, Yang EY, *et al.* Increased cell proliferation and decreased apoptosis characterize congenital cystic adenomatoid malformation of the lung. *J Pediatr Surg* 1998; 33: 1043–1046.
- 11 Wilson RD, Hedrick HL, Liechty KW, *et al.* Cystic adenomatoid malformation of the lung: review of genetics, prenatal diagnosis, and *in utero* treatment. *Am J Med Genet A* 2006; 140: 151–155.
- 12 Kotecha S, Barbato A, Bush A, *et al.* Antenatal and postnatal management of congenital cystic adenomatoid malformation. *Paediatr Respir Rev* 2012; 13: 162–170.
- 13 Werner HA, Pirie GE, Nadel HR, *et al.* Lung volumes, mechanics, and perfusion after pulmonary resection in infancy. *J Thorac Cardiovasc Surg* 1993; 105: 737–742.
- 14 Komori K, Kamagata S, Hirobe S, *et al.* Radionuclide imaging study of long-term pulmonary function after lobectomy in children with congenital cystic lung disease. *J Pediatr Surg* 2009; 44: 2096–2100.
- 15 Downard CD, Calkins CM, Williams RF, *et al.* Treatment of congenital pulmonary airway malformations: a systematic review from the APSA outcomes and evidence based practice committee. *Pediatr Surg Int* 2017; 33: 939–953.
- 16 Keijzer R, Chiu PP, Ratjen F, *et al.* Pulmonary function after early vs late lobectomy during childhood: a preliminary study. *J Pediatr Surg* 2009; 44: 893–895.
- 17 Hijkoop A, van Schoonhoven MM, van Rosmalen J, *et al.* Lung function, exercise tolerance, and physical growth of children with congenital lung malformations at 8 years of age. *Pediatr Pulmonol* 2019; 54: 1326–1334.
- 18 Harris M, Clark J, Coote N, *et al.* British Thoracic Society guidelines for the management of community acquired pneumonia in children: update 2011. *Thorax* 2011; 66: Suppl. 2, ii1–ii23.
- 19 Patradoon-Ho P, Fitzgerald DA. Lung abscess in children. *Paediatr Respir Rev* 2007; 8: 77–84.