

Respiratory distress in a newborn

P Ip, KL Chan, H Ngan, US Khoo

Presentation of case

Dr KL Chan*: A 2-day-old girl presented with respiratory distress. She was born at 40 weeks gestation by spontaneous vaginal delivery, and had a birth weight of 3670 g. The Apgar scores were 8 at 1 minute and 10 at 5 minutes. Would any of the students like to tell us what the Apgar score is?

Student: It is a scoring system used to evaluate the general condition of a newborn baby. The criteria are based on heart rate, respiratory effort, muscle tone, response to stimuli, and skin colour.

Dr Chan: The score was introduced by an American anaesthetist, Dr Virginia Apgar. A score between 8 and 10 indicates a baby in excellent condition, whereas a score of below 7 is cause for concern. The score also reflects the progress of resuscitation and can give a prognostic indication. So this patient was in good condition at birth. Physical examination revealed a healthy-looking baby with no dysmorphic features. Her skin colour was pink. However, she was in respiratory distress, with a respiratory rate of 70 per minute. On examination of the chest, there was decreased air entry on the right. It was resonant on percussion and occasional crepitations were heard on auscultation. No bowel sound, however, was audible in the chest.

Cardiovascular examination revealed normal heart sounds with no murmurs. Other systems were unremarkable. The initial blood biochemistry and haematology results were normal. However, the arterial blood gases initially done when the baby was breathing room air showed respiratory acidosis, with a pH of 7.14. The $p\text{CO}_2$ was 10.3 kPa (normal range, 4.4 - 5.9 kPa), $p\text{O}_2$ was 4.1 kPa (normal range, 10.0 - 14.0 kPa) and the base excess was -5.5 mmol/L (normal range, -10.0 - -2.0 mmol/L). Hypoxia and hypercapnia were evident.

Differential diagnoses

Dr Chan: The likely causes of respiratory distress in a newborn include pneumonia—either congenital or due to meconium aspiration, conditions where there is an air leak, such as pneumothorax, pneumomediastinum, and pulmonary interstitial emphysema, pulmonary dysplasia, diaphragmatic hernia, lobar emphysema, respiratory distress syndrome (RDS) and congenital cystic adenomatoid malformation. Other non-respiratory system-related causes include congenital heart diseases (e.g. transposition of the great vessels), heart failure, anaemia, hypovolaemia, metabolic acidosis, congenital neuromuscular disease, and increased intracranial pressure.

Prof H Saing†: Since there are doctors from the Ear, Nose and Throat department (ENT) here, would Dr Hui like to list the ENT causes of respiratory distress for the benefit of the students?

Dr Y Hui‡: The possible ENT causes can be divided into those resulting from extrathoracic or intrathoracic airway obstruction. Extrathoracic airway obstruction causes include include choanal atresia, adenoid hyperplasia, retroplated tongue—as in Pierre-Robin syndrome, nasopharyngeal teratoma, sublingual dermoid, lingual thyroid, congenital sibglottic stenosis, subglottic haemangioma, and tracheal malacia. Intrathoracic obstruction causes include cystic hygroma, duplication cyst of bronchus, oesophageal atresia with tracheo-oesophageal fistula, the presence of vascular rings and related anomalies, mediastinal teratoma, and bronchial malacia.

Dr Chan: The chest and abdominal X-ray were taken. Would Professor Ngan like to give us some comments?

Prof H Ngan§: The plain X-ray (Fig 1) showed large cystic spaces in the right chest, displacing the mediastinum to the left, and the left lung was compressed. Air-filled bowel loops were present in the abdomen, but they did not run towards the right dia-

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phragm whereas the air-filled cystic spaces were present in the right chest. These observations exclude the diagnosis of a diaphragmatic hernia.



Fig 1. Plain radiograph of the chest showing mediastinal shift to the left and multiple large air-filled cystic spaces on the right



Fig 2. CT scan showing multiple cystic spaces in the right hemithorax with some normal lung tissue at the upper lobe

Dr Chan: A computed tomography (CT) scan of the thorax was also taken.

Prof Ngan: The CT scan (Fig 2) showed large air-filled cystic spaces in the right chest with some normal lung tissue at the upper lobe. These suggested a diagnosis of congenital cysts of the lung.



Fig 3. Photograph of the cross-section of the lobe of the lung showing multiple large cysts and the adjacent compressed lung

Dr Chan: The patient was given 30% oxygen via the headbox and her oxygen saturation (SaO_2) reached 85%. Her general condition was stable, with the acidosis gradually being corrected. Her pH was 7.41 on the following day. The blood gases showed a PCO_2 of 5.3 kPa, PO_2 of 6.6 kPa, and a base excess of +1.1 mmol/L, indicating that she was still slightly hypoxic. Because of the persistent respiratory distress and a definite lesion in the right chest, intraoperative exploration was indicated. A right thoracotomy was carried out at the fourth intercostal space. Intraoperatively, the right lower lobe was found to contain multiple cysts, with the largest one measuring 5 cm at its widest diameter, occupying nearly the whole of the right chest. A right lower lobectomy was performed.

Pathology

Dr P Ip¹¹: The specimen we received was a lobe of lung measuring 8.0 x 4.5 x 3.5 cm, with multiple cysts

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of varying sizes found internally, separated by complete and incomplete fibrous septae (Fig 3). The adjacent lung tissue was compressed. Histologically, the cysts were lined by ciliated, pseudostratified, columnar epithelium (Figs 4 and 5) and focal clusters of mucous-secreting cells were seen (Fig 6). The smaller cysts were lined by columnar- to cuboidal-type epithelium. The walls of the cysts contained thick fibromuscular and elastic tissue (Fig 7), with focal cartilage plates. The final diagnosis was consistent with Type I congenital cystic adenomatoid malformation (CCAM).

Pathological discussion

Dr Ip: Congenital cystic adenomatoid malformation was first described by Ch'in et al in 1949 as a hamartomatous lesion.¹ There is unilateral involvement in 95% of the cases, either lung being affected with equal frequency. Ninety per cent of cases involve a single lobe, of which 60% are of the lower lobe.

Prof Saing: There are three clinicopathological types of CCAM, would you mind elaborating more about the different types?

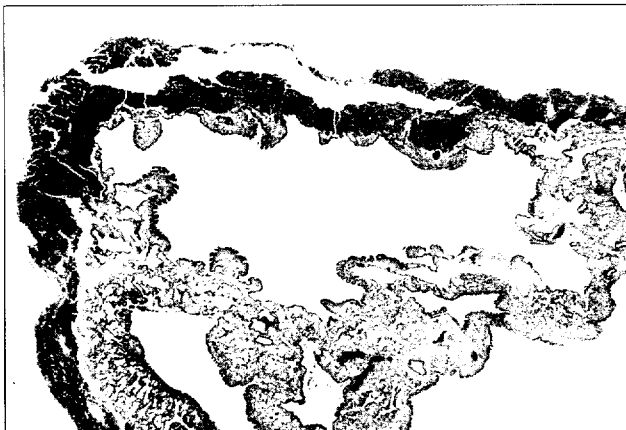


Fig 4. Photomicrograph showing one of the larger cysts, with adjacent lung parenchyma compressed (H&E, x 100)

Dr Ip: Congenital cystic adenomatoid malformation has been classified into three types, based on clinicopathological features.² Type I (Fig 8) is usually diagnosed in patients aged from 1 week to 20 years. It consists of multiple large cysts 3 to 10 cm in size, separated by normal alveoli. The walls of the cysts are usually composed of thick smooth muscle, elastic tissue, and cartilage, and are covered by respiratory type and cuboidal epithelium. Focally, mucous-producing cells are also found in 30% of cases. Type II (Fig 9) is usually diagnosed in patients aged up to 1 year, and consists

of medium-sized cysts 0.5 - 2.0 cm, resembling distended terminal bronchioles, separated by large alveoli-like structures. By alveoli-like structures, I mean they resemble the immature alveoli seen in foetal lungs at the pseudoglandular stage, when they are lined by cuboidal-type epithelium. Unlike Type I CCAM, the cyst wall in type II consists of a thinner fibrous wall devoid of smooth muscle or elastic fibres, and is covered by columnar epithelium. No mucous cells or cartilage are found. Type III CCAM (Fig 10) is usually diagnosed in neonates and infants up to the age of 1 month, consisting of numerous small cysts less than 0.5 cm in size. Histologically, the small cysts are alveoli-like structures that morphologically resemble immature foetal lung. In between the cysts are dilated bronchiolar structures, which have no direct communication with the tracheo-bronchial tree.³



Fig 5. Photomicrograph of the wall of a large cyst showing thick smooth muscle covered by ciliated pseudostratified columnar epithelium (H&E, x 300)

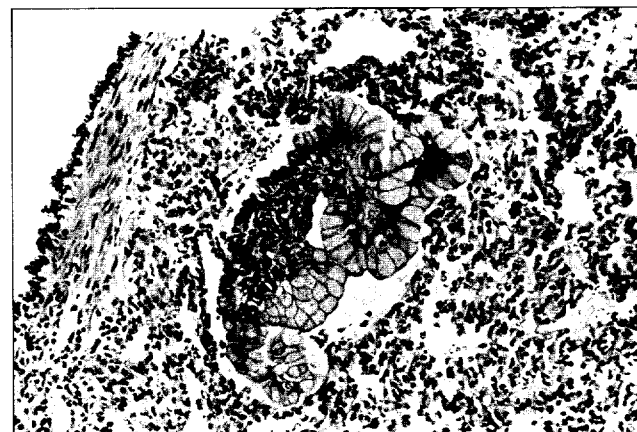


Fig 6. Photomicrograph showing focal cluster of mucinous cells (H&E, x 500)

Prof Wong¹: The cysts of the lung appear similar to those found in the kidneys, whether they are type I, II,

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or III. Is there any association of this disorder with polycystic kidney disease?

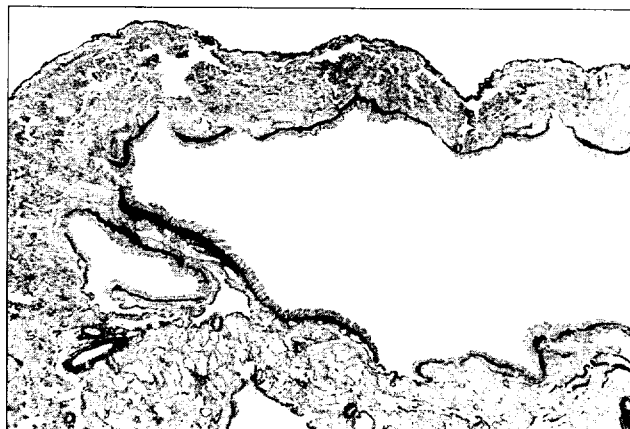


Fig 7. Photomicrograph of cyst wall showing the presence of abundant elastic fibres (Elastic van Gieson, x 200)

Dr Ip: There has been no case reported with this association. However, co-existing renal lesions with CCAM include bilateral renal agenesis and the prune-belly syndrome.^{2,4} Other associated anomalies more frequently reported include extralobar pulmonary sequestration, pulmonary hypoplasia, cardiovascular malformations, diaphragmatic hernia, hydrocephalus, macrocephaly, myelomeningocele, and jejunal atresia. Most of these anomalies were found in patients with Type II CCAM, and are found in Type I less frequently. There has been no association of these anomalies with Type III CCAM.

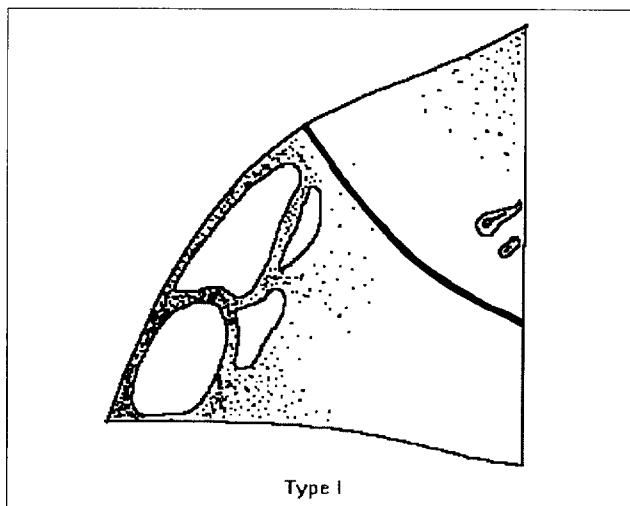


Fig 8. Drawing illustrating Type I CCAM with multiple large cysts (3.0 - 10.0 cm at widest diameter)

Student: What is the clinical outcome, prognosis, and histogenesis of CCAM?

Dr Ip: In many cases, patients with CCAM either die in-utero, or die neonatally due to pulmonary hypoplasia.⁵ In cases of Type I CCAM, patients can be asymptomatic at birth, and remain well until early adulthood, when they present with recurrent pneumonia.^{6,7} A few cases have been reported of patients with a history of CCAM who developed rhabdomyosarcoma and bronchioloalveolar carcinoma of the lung in later life.^{8,9}

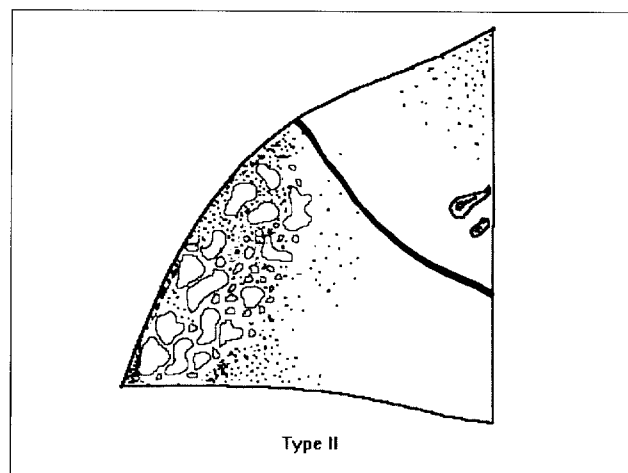


Fig 9. Drawing illustrating Type II CCAM with numerous smaller cysts (0.5 - 2.0 cm at widest diameter) separated by multiple alveoli-like structures

Since the number of reported cases are too few, there has been no published data on prognosis. However, Neilson et al reviewed 50 cases and found that maternal polyhydramnios, foetal ascites, and mediastinal shift are associated with a poorer prognosis.⁵ They also found that type I CCAM has the highest survival rate. Types II and III have similar survival rates of approximately 50%. One would expect Type II to have a rate between those of Types I and III, but this finding can be accounted for by the high frequency of associated anomalies.

Moerman et al propose a theory on the pathogenesis of CCAM.³ They suggest that a lack of communication between the lesion and the tracheo-bronchial tree is responsible, i.e. an absent or atretic segmental bronchus, and suggest that it represents a developmental arrest of the endodermal tracheo-bronchial bud, or its branching process. Thus, the end result is bronchial atresia. They also suggest that if growth proceeds beyond the atretic segment, an abnormality will arise. The type of malformation (I, II, or III) is determined by the extent of dysplastic lung growth beyond this atretic segment. They conclude that CCAM is either a primary defect in organogenesis, or is a developmen-

tal defect secondary to a vascular abnormality supplying the lung.

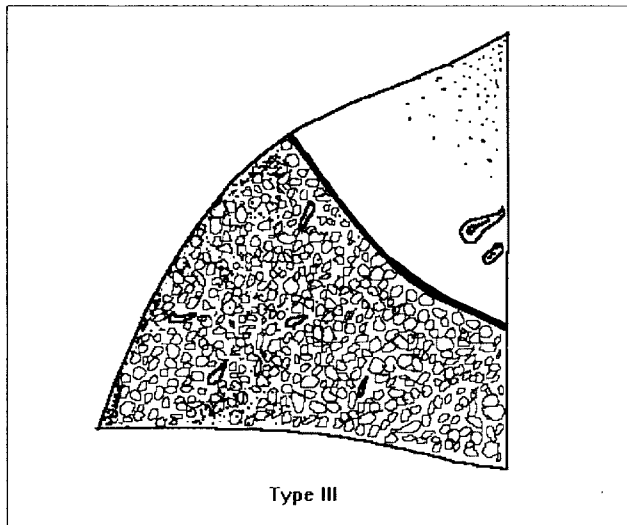


Fig 10. Drawing illustrating Type III CCAM with multiple small alveoli-like cysts (< 0.2 cm at widest diameter) involving an entire lobe separated by regularly spaced bronchiolar-like structures

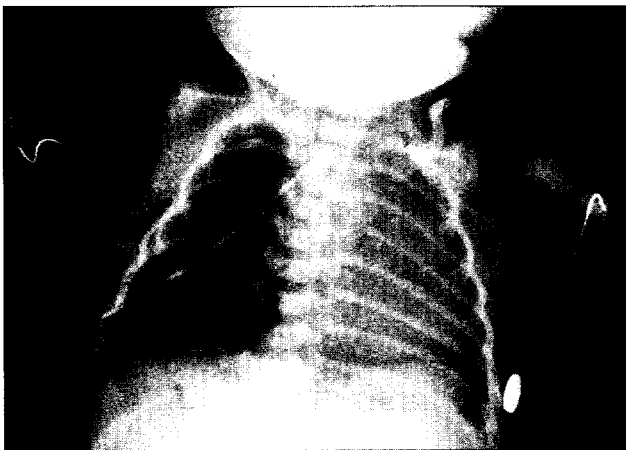


Fig 11. Plain radiograph of the chest after operation showing a right chest drain in-situ and the remaining right lung occupying two-thirds of the hemithorax. Pneumothorax is still present.

Dr Chan: The post-operative period was uneventful. A post-operative chest X-ray was taken.

Prof Ngan: The chest X-ray (Fig 11) showed a chest drain in the right chest with the remaining right lung occupying two-thirds of the hemithorax. Pneumothorax was still present.

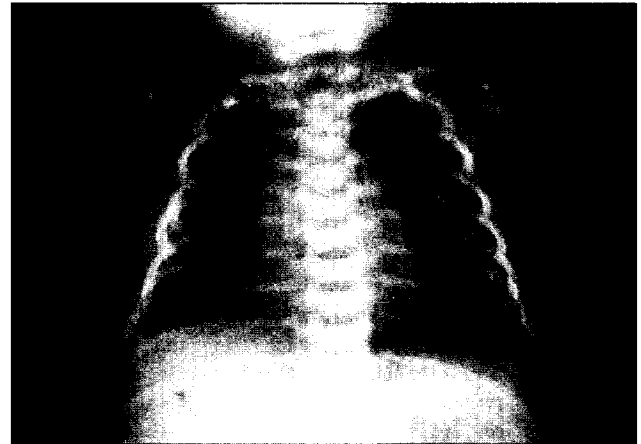


Fig 12. Plain radiograph of the chest before discharge showing full expansion of the remaining right lung with disappearance of the pneumothorax

Dr Chan: The child was weaned off the ventilator on post-operative day 2, the chest drain was removed on day 5, and she was discharged on day 16.

Prof Ngan: The chest X-ray (Fig 12) showed full expansion of the remaining right lung, with disappearance of the pneumothorax.

Dr TG Lorentz^{**}: In cases when there is severe dyspnoea and where specialist care is not available, do you think that inserting a catheter in the chest can help to release the pressure?

Prof Saing: It is not advisable because of the danger of inducing a broncho-cutaneous fistula.

Dr Chan: The antenatal use of ultrasound-guided pleuro-amniotic catheter insertion in-utero has been attempted to release the tension of the large cysts, so that the pressure effect on the normal lung can be reduced, thus improving the survival of those foetuses with huge cysts.¹⁰ Furthermore, with the use of ultrasound, prenatal diagnosis can identify those babies that should be delivered at a tertiary centre with expertise in managing these lesions surgically, immediately after birth.

Pathological diagnosis:

Type I congenital cystic adenomatoid malformation of the lung

Acknowledgements

The authors would like to thank the other participants of the meeting.

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PICTORIAL MEDICINE



This is the retinal appearance of a 20-year-old man who presented with a total white cell count of $341 \times 10^9/L$ and a platelet count of $29 \times 10^9/L$. He was subsequently diagnosed as having acute biphenotypic leukaemia. He complained of reduced visual acuity and fundoscopic examination showed multiple retinal haemorrhages and Roth's spot. The latter are thought to be due to leucostasis causing central ischaemia with surrounding haemorrhages.

Submitted by W Yeo, N Wickham, PJ Johnson.

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