

**Case report:**

**Congenital Cystic Adenomatoid Malformation  
(CCAM): Two Rare Cases Report in Sudanese  
Children**

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**Abstract:**

*We here report a rare case of congenital cystic adenomatoid malformation [CCAM]. The first case presented early in the neonatal period with unilateral lung cysts and have favorable outcome. The second case present at 2months of age with bilateral congenital lung cyst.*

**Key words:** Congenital cystic adenomatoid malformations (CCAM), Sudanese children

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## **CASE REPORT (1):**

An Eleven days male baby delivered at term by vaginal delivery at home. He cried immediately and breast fed same day. He was of average weight. The baby developed shortness of breath on the 11<sup>th</sup> day associated with severe respiratory distress. His mother was 24 years old and had malaria at 5<sup>th</sup> month of pregnancy with treatment. The elder two siblings were died, first at 7 years of age following accidental head trauma and the second at early infancy with diarrhoeal disease.

On examination: he was ill with severe respiratory distress. He was afebrile. His vitals were as follows: RR 78/min, HR 136/min, temperature 36.2C and oxygen saturation 88% at room air. His weight and length were average. Chest examination showed signs of respiratory distress with decreased air entry bilaterally but the right side < left side without any added sounds. Immediately, the baby admitted to HDU and put on O<sub>2</sub>, antibiotics, NPO and IV fluids.

His CBC and renal function tests were normal. Blood film for malaria was negative. Chest X-ray (case1) showed. CT scan (case 1) of the chest showed right lower lobe cystic adenomatoid malformation (CCAM), mediastinal shift to the left side and decreased lung volume in the left side (90% ↓). The baby underwent surgical operation and connected to mechanical ventilator for 8 days. (case 1 post operative photo & x ray).

## **CASE REPORT (2):**

A 2 months old male baby delivered by SVD at term at hospital but cried after a while and discharged home after 2 hours. 3 weeks ago he developed cough, shortness of breath and high grade fever with hospital admission for 8 days and minimal improvement then referred to our hospital with fever and

respiratory distress. He is the first baby for a consanguineous parents. His mother is 20 years old and father is 32 years. The pregnancy complicated by fever and skin rash in the last trimester. He is not vaccinated.

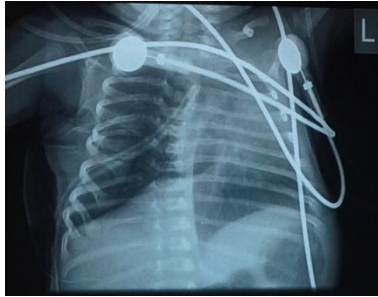
On examination: he was ill with respiratory distress (case 2) and febrile. His vitals were as follows: RR 60/min, PR 108/min, temperature 37,9C and oxygen saturation 85% at room air. His length and head circumference are within normal limits but weight is below average. Chest examination showed signs of respiratory distress with decreased air entry in the right side of the chest with no added sounds. The baby admitted to HDU and put on O<sub>2</sub>, antibiotics, antipyretics and IV fluids with minimal improvement.

The CBC showed Hb of 10 gm/dl, TWBC 27,000 and platelets 355,000. His renal function tests and electrolytes are normal. Blood Film For Malaria (BFFM) is negative. Chest X-ray (case 2) showed right side opacity involving all the Rt lung field. CT scan chest (case 2) revealed bilateral cystic adenomatoid malformation (CCAM) mostly type I on the right side with bilateral superadded infection.

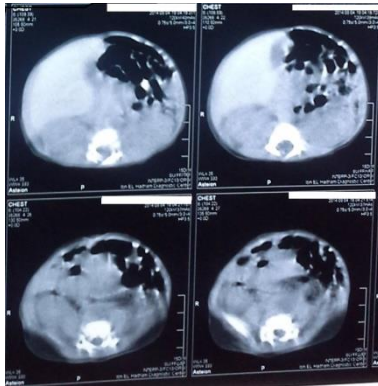
### **Case 1: Chest x ray (pre-operative)**



**Case 1: chest x ray (post operative)**



**Case 1: CT scan chest**



**Case 1: post-operative with chest tube**



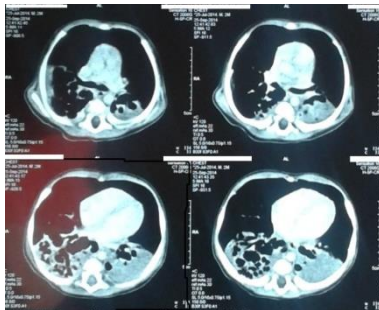
**Case (2): sub-costal recession**



**Case 2: Chest X-ray**



**Case 2: Chest CT scan**



## DISCUSSION

Congenital pulmonary airway malformation (CPAM), previously known as congenital cystic adenomatoid malformation (CCAM), is a rare developmental anomaly of the lower respiratory tract [ 1-5 ]. Affected patients may present with respiratory distress in the newborn period or may remain asymptomatic until later in life. Many cases are now detected by routine prenatal ultrasound examination.

Data from large population registries suggest an incidence of congenital lung cysts in the range of 1 per 8300 to 35,000 live births [ 5 ]. Large-cyst subtypes account for about 70 percent of CPAMs, or 2 to 8 per 100,000 live births.

CPAMs occur sporadically. Their formation is not related to maternal factors such as race, age, or exposures. In some series, lesions that present in infancy have a slight male preponderance [6-9], although others found no gender predilection [10-11]. Congenital cystic adenomatoid malformations were divided into three major types based upon the size of the cysts and their cellular characteristics (predominantly bronchial, bronchiolar, or bronchiolar/alveolar duct cells)[1.2.12]. Under this classification scheme, more than 65 percent of CPAMs were type 1, type 2 comprised 20 to 25 percent, and type 3 comprised 8 percent [ 1,2 ].

In the present classification scheme these congenital cystic lung lesions are now called congenital pulmonary airway malformations (CPAM), and two additional types (0 and 4) were added. Type 0 arises from the trachea, and type 4 lesions have alveolar/distal acinar origins [13-15]. Each type of CPAM has distinct pathologic characteristics [12-15].

The presentation of congenital pulmonary airway malformation (CPAM) is variable. Many are identified by routine prenatal ultrasound examination. Affected newborns

can present with signs and symptoms of respiratory distress or be asymptomatic.

CPAMs presenting prenatally are classified by their ultrasound findings and gross anatomy [16,17]. Microcystic lesions consist of cysts <5 mm in diameter and appear echogenic and solid, while macrocystic lesions contain one or more cysts >5 mm in diameter.

Prenatal magnetic resonance imaging (MRI) may help distinguish CPAM from congenital diaphragmatic hernia (CDH) or other less common lesions. In one series, 16 of 18 patients with fetal lung masses were diagnosed as CPAM by ultrasound [18]. However, MRI imaging confirmed nine of the lesions to be CPAM, three were CDH, two were bronchopulmonary sequestrations (BPS), and one each was a foregut cyst, lung atresia, tracheal atresia, and bronchial stenosis.

Poor prognostic signs include large CPAM size, bilateral lung involvement, polyhydramnios, and mediastinal shift [19]. However, there are no reliable criteria for determining which lesions will grow and result in hydrops versus those that will stabilize or regress.

Approximately 25 percent of infants with CPAM detected prenatally also have other structural anomalies; most of these cases are associated with type 2 CPAM [20]. In patients with such anomalies, a fetal karyotype should be obtained, although CPAMs typically are not associated with chromosomal abnormalities.

The management of congenital pulmonary airway malformation (CPAM) depends on whether the patient has respiratory distress or is asymptomatic.

In symptomatic patients, CPAM is treated by surgical resection [12,21-25]. Resection is frequently required in newborns with significant respiratory distress, but is often done electively in older children who present with less acute

symptoms. In older children, resection is usually performed to prevent recurrent infection and to eliminate concerns regarding malignancy.

In asymptomatic patients, surgical resection for all patients with bilateral or multifocal cysts or a family history of pleuropulmonary blastoma (PPB)-associated conditions is recommended. Also suggest surgical resection of lung cysts in infants or children without these features is suggested, but surveillance with serial CT scans is a reasonable alternative.[ 26 ]

The most common complication of congenital pulmonary airway malformation (CPAM) is recurrent pulmonary infection. In some cases, this may be the presenting sign. A rare complication is spontaneous hemopneumothorax [27]. The development of malignancy in some patients with previously unrecognized CPAM, while rare, is another reason for the recommendation to resect the lesions, even in asymptomatic patients.

We here reported two infant with CCAM who behaves like chronic lung disease. To the best of our knowledge, this is the second report from Sudan as the first reported case was done by Sirageldin MK Abdel Rahman et al 2013.[ 28 ]

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