

## Congenital Pulmonary Airway Malformation (CPAM) follow up from 23 week of gestation

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### ABSTRACT

The development of the respiratory system starts at 3 weeks of gestation, and aberrations in developmental processes may result in structural abnormalities collectively referred to as bronchopulmonary foregut malformations. These lesions include congenital cystic adenomatoid malformations (CCAMs), sequestrations and infantile lobar emphysema. Case presented is of right lung CCAM diagnosed at 23 weeks of gestation, followed during antenatal periods for complications, delivered at 39 weeks, planned thoracotomy with lobectomy done on 3<sup>rd</sup> day of life and followed up till 1 year.

Detailed anomaly scan and close monitoring for structural abnormalities of fetus is needed for appropriate management. Relationship of CPAM with early pregnancy severe infections remains to be established.

**Keywords:** Anomaly scan, CPAM, Lobectomy

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### INTRODUCTION:

The respiratory system starts its development from 3<sup>rd</sup> week of gestation, and aberrations in this development may result in structural abnormalities collectively referred to as bronchopulmonary foregut malformations. These lesions include congenital pulmonary airway malformations (CPAMs), sequestrations and infantile lobar emphysema. It is due to

improved antenatal ultrasounds that most CPAMs are diagnosed during antenatal period usually around the time of anomaly scan or can be identified in the neonatal period. Only few cases (10%) present after the first year of life usually after getting complications like infections<sup>1</sup> or respiratory problems. Congenital pulmonary airway malformation (CPAM) is a rare developmental abnormality of the lower respiratory tract firstly described as a distinct anomaly by Ch'in and Tang in 1949.<sup>2</sup> Many cases are now detected by routine prenatal ultrasound examination. Incidence reported is between 1:11,000 and 1: 35,000 live births.<sup>3</sup> As most case reported are found from tertiary care referrals so exact incidence in general population is not exactly found.

### CASE REPORT:

A case of type I CCAM is reported in which a large cyst with multiple out pouches was present in right middle lobe as a whole and lower lobe partially 35 years old female, married to a non consanguineous relationship was second gravida and had emergency caesarean section in her first pregnancy a year back because of fetal distress. There was no family history of any congenital disorder. It was a spontaneous conception and folic acid was commenced since start of pregnancy. Pregnancy was complicated by severe urinary tract infection during 6th and 7th week of gestation. *Klebsiella* was responsible organism and UTI was associated with abnormal liver function tests (raised transaminases and  $\psi$  glutamyl transpeptidase). It was treated with intravenous antibiotics. Pregnancy progressed well until 23 weeks when CPAM diagnosed during anomaly scan (Fig. 1).



**Figure 1 : Congenital pulmonary airway malformation of right lung at 23 weeks**

Detailed anomaly scan revealed it as isolated malformation with mild polyhydramnios. Left lung was normal. Fetal echocardiography was normal except mild left axis deviation. No other pregnancy complication was found and follow up planned with two weekly ultrasounds for cyst size, maximum vertical pool of liquor, growth scan, umbilical artery Doppler and any evidence of fetal hydrops. Cyst size didn't show any change (5.2×2.3 cm) so did polyhydramnios (Fig. 2).



**Figure 2 : Ultrasound at 32 weeks**

Multidisciplinary team approach with consultant obstetrician, ultrasonologist, neonatologist and pediatric surgeon was adopted. Couple opted for planned repeat caesarean section so elective c-section done at 39 weeks. Girl weighing 2.89 kg delivered with APGAR 5, 8 at 1 and 5 minutes. There was no respiratory problem in baby while observed in N.I.C.U. Planned thorocotomy done on third day of life so as to avoid risk of infection or respiratory problem in the neonate ( Fig. 3).



**Figure 3 : CT scan before surgery**

Complete middle and partial lower lobectomy done on right lung with a good recovery. Baby discharged on 7<sup>th</sup> post op day with regular fortnightly follow up which showed rapid and uneventful recovery and good air entry on right side of chest. Histopathological examination of the tissue removed showed lung tissue with large cystic spaces lined by ciliated pseudo columnar bronchial epithelium, surrounding alveoli dilated and lined by columnar to cuboidal epithelium and look like glands. Six monthly and one year follow up was normal with no respiratory problem.

### DISCUSSION:

CCAM is usually unilateral and located in one lobe only. Miller et al. have reported higher rates of involvement of lower lobes (right or left) and lower rates for middle lobe.<sup>4</sup> Majority of antenatally diagnosed CCAMs are left-sided.<sup>5,6</sup> Studies mentioned both gender contributions but males in most studies being more commonly affected.<sup>7</sup>

There is few data on bilateral CCAMs.<sup>4</sup> The significance of bilateral lesions may relate to a genetic predisposition to subsequent malignant change within the CCAM or elsewhere in the lung. One of the potential genes which can result this development disorder is HOXB5, as its expression is maintained at a level typical for early lung development.<sup>8</sup>

The accuracy of antenatal diagnosis is paramount in the care of the fetus with a CPAM. Prenatal ultrasound and MRI are used to establish its location of the lung, appearance, its blood supply and venous drainage by Doppler ultrasound and any displacements of other lung lobes in thoracic position, mediastinum, and the cardiac structures. CCAMs are fed by the pulmonary system (arterial and venous blood flow). Additional survey should include amniotic fluid volume, Doppler flow patterns in umbilical artery and ductus venosus, and placental thickness. There is no known association of CCAM with chromosome aberrations, but additional structural abnormalities in the renal, cardiac, skeletal and gastrointestinal systems should be assessed as these can be found in 10 to 20% of cases.<sup>9</sup>

CPAM monitoring requires serial ultrasounds every fortnightly. Ultrasound assessment should include change in cyst size, polyhydramnios and hydrops fetalis as these are predictors of poor outcome. Macrocystic (type I) disease changes little by the end of pregnancy, whereas microcystic (type II) has tendency to shrink.<sup>7</sup>

Fetal intervention can be based upon gestational age, the size of the lesion, the mother's health and the development of fetal hydrops. In utero techniques include open maternal-fetal therapy with fetal thoracotomy and lobectomy, thoracoamniotic shunting of macrocystic CCAM's and third trimester EXIT delivery with fetal thoracotomy, and lobectomy on maternal placental bypass.<sup>10</sup> These interventional techniques are available only at limited centers.

As the lesions usually regress toward the end of the second trimester or the beginning of the third trimester, pre-term delivery is usually not indicated. Prematurity should be avoided so as to

minimize the risk of complications for neonate. Vaginal delivery is of choice in most of these lung lesions. Delivery should be conducted in a setup which has immediate access to a tertiary neonatal intensive care unit.

Most of the neonates with CPAM are asymptomatic. Presentation can be with respiratory difficulty or overt respiratory failure. Infants and young children with borderline symptoms may become overtly symptomatic or may persist with tachypnea, feeding difficulties and failure to thrive. A chest radiograph may reveal a localized lesion. Main role of CT is to differentiate these cysts from other lung lesions like bronchogenic cysts, lobar emphysema, and from sequestration, and accurately localize the site of lesion for surgical planning.

Elective surgery is associated with improved outcomes especially when performed in asymptomatic infants it results in less morbidity<sup>11</sup> Simple resection of the involved tissue is enough but lobectomy can eliminate the risk of retained lesions which needs follow-up.<sup>12</sup>

Appropriate timing of cyst removal is not clear but if surgery is elected it should be performed in the first 10 months of life.<sup>13</sup> Expectant management can be considered.

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