# CHAOS: Prenatal imaging findings with post mortem contrast radiographic correlation

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ABSTRACT

Congenital high airway obstruction syndrome is a rare fetal anomaly with characteristic constellation of prenatal findings on ultrasound and MRI. The typical triad of imaging features are enlarged and echogenic lungs, flattening or inversion of diaphragm and fetal hydrops. Early prenatal recognition of congenital high airway obstruction syndrome by ultrasound and/or MRI is mandatory for the appropriate perinatal management. We report a case of a male fetus with typical imaging findings of congenital high airway obstruction syndrome on ultrasound and MRI at 19 weeks of gestation. The role of contrast radiographs of fetal airways, including retrograde laryngogram, in confirming the postnatal diagnosis of this fetal condition is demonstrated. The prenatal imaging findings were correlated with contrast radiographs of upper airways, sonography of aborted fetus and fetal autopsy findings.



## CASE REPORT

### CASE REPORT

A 32 year old woman (gravida 4, para 1, abortion 3, live 1) was referred to our department for a routine prenatal scan at 19 weeks of gestation. There was no history of consanguinity. Ultrasound showed enlarged and echogenic lungs bilaterally with inversion of diaphragm [Fig 1a, 1b]. Fetal heart was compressed and centrally positioned in the thorax [Fig 2a, 2b]. Fetal ascites was present [Fig 3]. Additional findings of perinephric fluid collection, subdural collection and subcutaneous edema were noted confirming hydrops fetalis [Fig 4]. Together, all signs on ultrasound provided the direct evidence of congenital high airway obstruction syndrome (CHAOS) of the fetus. Detailed evaluation on ultrasound revealed significantly dilated and fluid filled tracheobronchial tree [Fig 1a, 5]. No additional fetal abnormalities were noted. Previous antenatal scans done at 12 weeks and 6 weeks of gestational age were unremarkable. Fetal MRI was performed to evaluate the dilatation of tracheobronchial airway and confirm the level of obstruction. Fetal MRI confirmed the ultrasound findings of voluminous lungs with resultant inversion of diaphragm and demonstrated the presence of

abnormally dilated trachea and bronchi with sharp proximal cut off at the level of larynx [Fig 6, 7, 8]. The airway obstruction was confirmed at the level of larynx, with absence of any extrinsic lesion. Polyhydramnios and thickened placenta (measuring 38mm in maximum thickness) were present [Fig 1a, 7, 8]. A prenatal diagnosis of CHAOS with intrinsic obstruction at the level of larynx was made. The parents were counseled in detail about the condition. An option of ex utero intrapartum treatment (EXIT) procedure for near term management of fetal airway was proposed but was declined by the parents. In view of poor prognosis of the clinical condition, the parents chose to terminate the pregnancy. Medical termination was performed by labor induction using misoprostol at 20 weeks of gestation. The patient delivered a stillborn male child weighing 370 gram with gross distention of abdomen. Post mortem radiograph of the aborted fetus demonstrated opaque thorax with splaying of rib cage [Fig 9]. Contrast radiographic study of fetal airways was done post mortem to confirm the exact level of airway obstruction. 5ml of Iohexol (350mgI/ ml) was introduced into the oral cavity of the fetus with infant feeding tube. Antero-posterior and lateral radiographs of cervical region showed normal contrast filling of pharynx, with an abrupt cut off at the distal end of pharynx. There was lack of contrast filling distal to pharynx suggestive of obstruction at that level [Fig 10a, 10b]. Retrograde laryngogram was performed to assess the complete length and degree of stenosis by introducing contrast into the fetal upper airway in a retrograde manner via trachea. Fetal trachea was identified on ultrasound and another 5ml of iohexol was injected into the fetal upper airways via trachea under sonographic guidance. Trachea showed dilatation with abrupt proximal cut off. Review radiographs revealed a complete lack of contrast filling in the region of fetal larynx, giving an hour glass deformity on retrograde laryngogram [Fig 11a, 11b]. A diagnosis of complete laryngeal stenosis was made. A sharp border between occluded larynx and dilated tracheal column below was demonstrated [Fig 11a, 11b]. Ultrasound of fetal cervico-thoracic region demonstrated stenosis over a cephalo-caudal length of 5mm located proximal to trachea [Fig 12]. Fetal ascites, pleural effusion and dilatation of trachea were confirmed [Fig 13a, 13b]. Real time ultrasound revealed an additional finding of tracking of contrast into oblique, blind ending channels via pharynx raising a possibility of developing pharyngo-tracheal/ laryngo-tracheal communication channels [Fig 14]. Fetal laryngoscopy revealed normal vocal cords, with laryngeal stenosis noted distal to vocal cords, suggesting a possibility of subglottic laryngeal stenosis as the underlying cause of CHAOS. Fetal autopsy was done after taking a proper consent and revealed abnormal cricoid cartilaginous ring, hard on palpation and having elliptical shaped morphology (long axis oriented along the anteroposterior plane) and no perceivable lumen [Fig 15a, 15b]. A final diagnosis of CHAOS with congenital subglottic stenosis due to an elliptical cricoid cartilaginous ring was established.

## DISCUSSION

CHAOS is a rare, life threatening fetal condition, usually with a lethal outcome [1]. Prenatal sonography alone is sufficient to make a diagnosis of this syndrome.

*Etiology & Demographics:*

The true incidence of CHAOS is unknown [1,2]. The first report of prenatal diagnosis of CHAOS came in 1989 by Arizawa et al [1,3]. Literature search revealed at least 36 prenatally diagnosed cases till 2007 with few additional cases reported sporadically till date [1,3]. The term CHAOS was coined by Hedrick et al in 1994 [1,4]. CHAOS is caused by complete or near complete obstruction of upper airways in a fetus. The upper airway obstruction in CHAOS blocks the normal clearance of fluids produced by the lungs through the larynx, resulting in dilated and fluid filled tracheobronchial tree. Laryngeal atresia is the most frequent cause, other etiologies being trachea atresia, laryngeal or tracheal webs, subglottic stenosis or atresia, obstructing laryngeal cysts, and laryngeal/ tracheal agenesis [1,2,5]. Subglottic stenosis, the etiology of CHAOS in the present case, is a congenital anomaly of larynx with developmental defect in the supporting cricoid cartilage, thus resulting in intrinsic obstruction at this level. A unique subtype of congenital subglottic stenosis results from elliptical cartilaginous ring where cricoid cartilages are hard and elliptical in morphology [6]. Fetal autopsy in our case revealed the presence of elliptical shaped cricoid cartilage with its long axis oriented in anteroposterior plane. Genetic etiology of elliptical cricoid ring is unknown [7]. Embryologically, as per Frazer theory, the cricoid cartilage normally obliterates its lumen completely at seven weeks of gestation. A failure of adequate recanalization of the larynx at 9-week embryo stage results in subglottic stenosis [6,7]. Most cases of typical CHAOS are sporadic, however few cases in literature suggest autosomal dominance [8]. CHAOS may occur as a part of Fraser syndrome which is a rare, recessive congenital malformation characterized by cryptophthalmos, syndactyly and urogenital defects [9]. Other genetic syndromes associated with CHAOS are short rib polydactyl syndrome, VACTERL syndrome, chromosomal abnormalities such as deletion of chromosome 5p, partial trisomy 5 and partial trisomy 16q [8]. Antenatal diagnosis of CHAOS in a fetus thus entails genetic counseling and management of further pregnancies. The fetus in our case lacked any additional anomalies, thus excluding a syndromic association.

*Clinical & Imaging Findings:*

Diagnosis of CHAOS is usually made on ultrasound between 18 to 31 weeks of gestation, with a recent case reported as early as in 15 weeks of gestation with transvaginal sonogram [3]. The prenatal imaging diagnosis of CHAOS is inferred from secondary changes as enlarged and echogenic lungs, flattening or inversion of diaphragms, fetal ascites and dilated distal airways [1,5]. Findings of CHAOS on prenatal ultrasound indicate complete/near complete obstruction of upper airways. The congested lungs cause mediastinal compression and lead to increased intrathoracic pressure, resulting in impaired cardiac filling, fetal cardiac failure, placentomegaly, ascites and non-immune hydrops. The echogenic appearance of lungs on ultrasound is attributed to

the increased number of tissue fluid interfaces. Fetal heart assumes a more central position due to voluminous lungs. Because of a difficulty in swallowing amniotic fluid, most cases of CHAOS are associated with polyhydramnios, attributable to obstruction of esophageal clearance of amniotic fluid by the enlarging lung mass. Features of CHAOS on ultrasound are diagnostic, however fetal MRI provides better detail of the degree and location of airway obstruction [5,10]. With its large field of view and high soft tissue contrast, fetal MRI has an advantage over ultrasound in detailing exquisite definition of fetal anatomy and facilitating treatment plan [5,10]. Retrograde laryngogram is a contrast radiographic procedure for accurately confirming the level and degree of laryngo-tracheal obstruction. An interesting finding in our case was the oblique tracking of contrast into the blind ending channels posteriorly at the level of the pharynx, noted during contrast injection under ultrasound guidance. This can be explained by the presence of minor posterior fistulous communications, as also described in the past in laryngeal atresia [1]. Recent reports in literature have suggested spontaneous development of pharyngotracheal/ laryngotracheal channels resulting in regression of hydrops fetalis in some cases of CHAOS [10,11]. However, the presence of communication channels could not be confirmed on contrast radiographs in our case.

*Treatment & Prognosis:*

Early diagnosis of this condition guides the parents in decision making whether to terminate the pregnancy, opt for fetal delivery with ex-utero intrapartum treatment (EXIT) procedure or to continue the pregnancy with no fetal intervention [11,12]. In EXIT procedure, the fetus is partially delivered by cesarean section and undergoes intubation or surgical procedures while the placenta and umbilical cord remain intact. EXIT procedure allows for increased survival rates in fetuses with CHAOS, however, entails a significant risk of brain damage or permanent tracheostomy. Fetoscopic deobstruction is another form of fetal therapy in select subsets of upper airways obstruction [13]. Cases of CHAOS with normal larynx and isolated upper tracheal obstruction may benefit from fetoscopic surgery [13]. In emergency situation, neonatal tracheostomy is considered.

*Differential Diagnoses:*

1. Bilateral congenital cystic adenomatoid malformation (CCAM Type III) is a fetal lung condition that closely mimics CHAOS on imaging. CCAM is a main non airway obstructing lesion usually lobar, mostly left sided and does not involve the entire lung, hence will show a compressed lung rim, not seen in CHAOS [14]. Bilaterality in CCAM is very rare (2%). Fetal hydrops in CCAM is uncommon (5%). In CCAM, in sharp contrast to CHAOS, fluid flow can be demonstrated in trachea during fetal breathing [2,14]. Identification of dilated tracheobronchial tree is a diagnostic feature specific to CHAOS, and differentiates it from CCAM.

2. Pulmonary sequestration: Pulmonary sequestration appears as echogenic, homogenous lung mass with incidence being 90% in the left hemithorax [15]. Large lesions may compress the esophagus and thoracic veins and cause hydrops. Detection of a systemic artery from the aorta to the fetal lung lesion is a pathognomonic feature of fetal pulmonary sequestration. Bilaterality of this disease is rare though few cases have been described [15]. Key feature of tracheal enlargement which is specific to CHAOS is missing in pulmonary sequestration.

To conclude: CHAOS is a rare clinical fetal anomaly with characteristic imaging features on ultrasound and MRI. This condition without fetal/neonatal intervention is fatal, hence prenatal diagnosis is of vital importance. Prenatal MRI, due to multiplanariety and excellent spatial resolution, plays an important role in surgical planning through identification of the level of obstruction. Retrograde laryngogram helps in postnatal confirmation of this condition which is useful in counseling of the patient and management of such cases in the future.

## TEACHING POINT

Congenital high airway obstruction syndrome has remarkable and very constant features of congested lungs, inverted diaphragms and ascites on prenatal ultrasound and MRI. Identification of this syndrome on prenatal imaging is vital for prenatal/perinatal intervention of potentially amenable cases, thus emphasizing on the significance of multidisciplinary team involvement in the management of congenital high airway obstruction syndrome.

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FIGURES

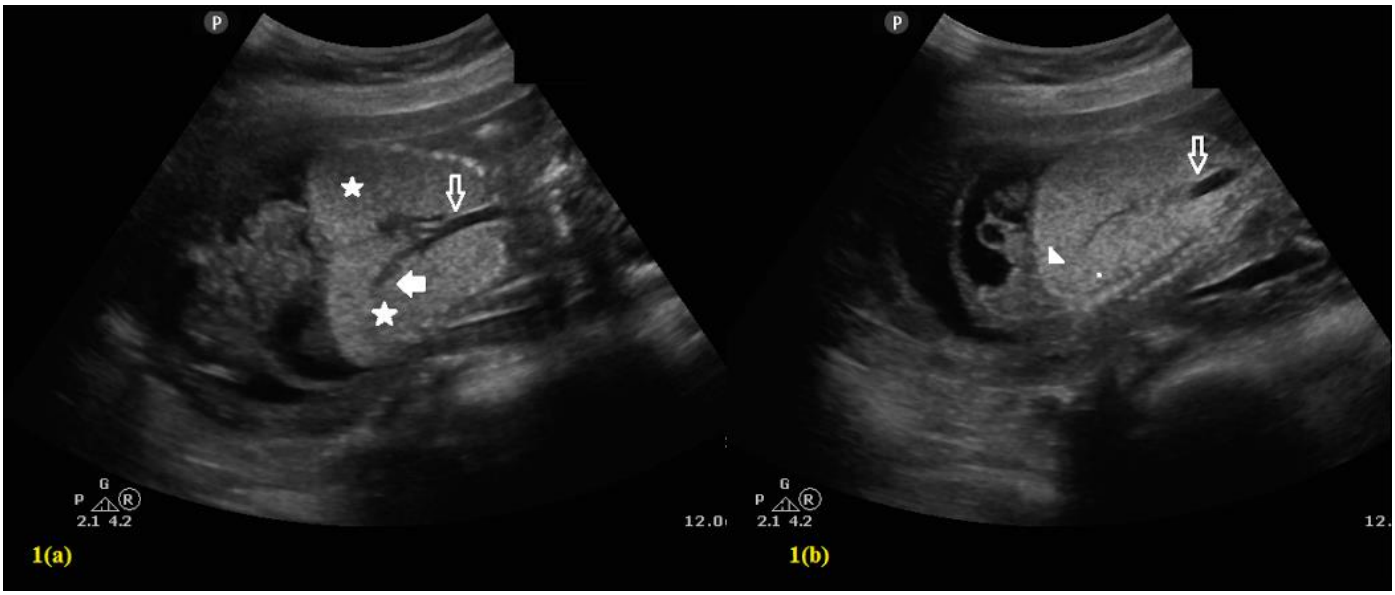



**Figure 1:** Male fetus at 19 weeks of gestation with congenital high airway obstruction syndrome. Findings: Prenatal sonographic image of fetal thorax in (a) coronal and (b) sagittal planes demonstrating echogenic lungs (asterisks) and dilated tracheal column (arrow) with abrupt proximal cut off. Bold arrowhead in Fig. 1a demonstrates dilated bronchus. Inversion of diaphragms is evident (arrowhead in Fig. 1b). Technique: Clearvue 650, Philips, Convex volume transducer (2-5 MHz).

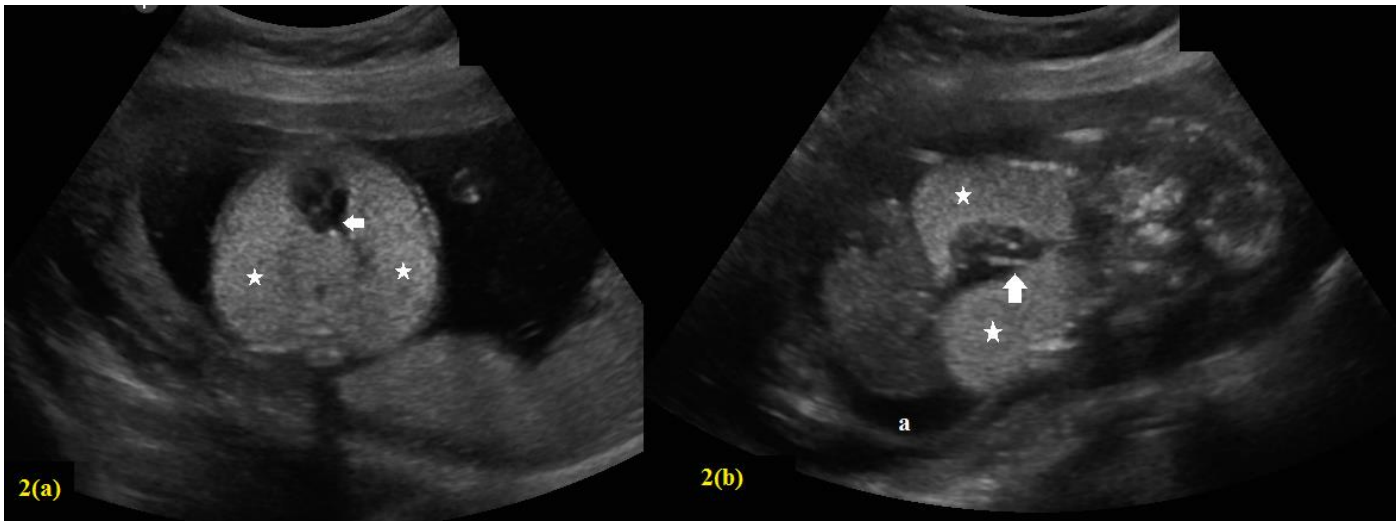



**Figure 2:** Male fetus at 19 weeks of gestation with congenital high airway obstruction syndrome: Prenatal sonographic image of fetal thorax in (a) axial and (b) coronal planes demonstrating compressed heart and pointing to the midline (bold arrowhead). Asterisks denote congested and echogenic lungs. Fetal ascites (denoted by a in Fig. 2b) is noted. Technique: Clearvue 650, Philips, Convex volume transducer (2-5 MHz).



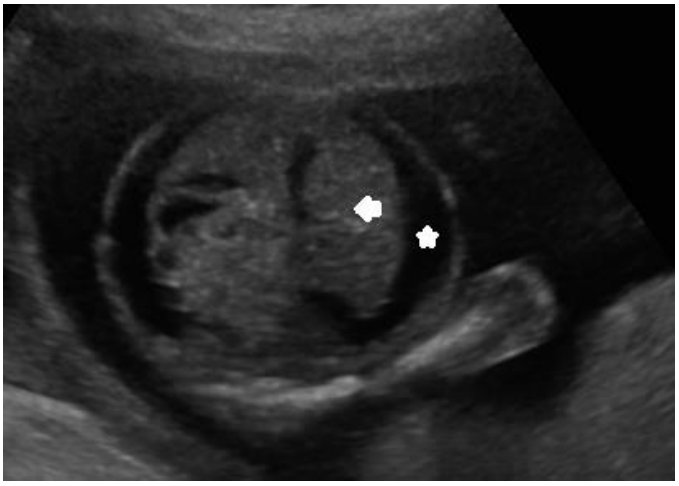
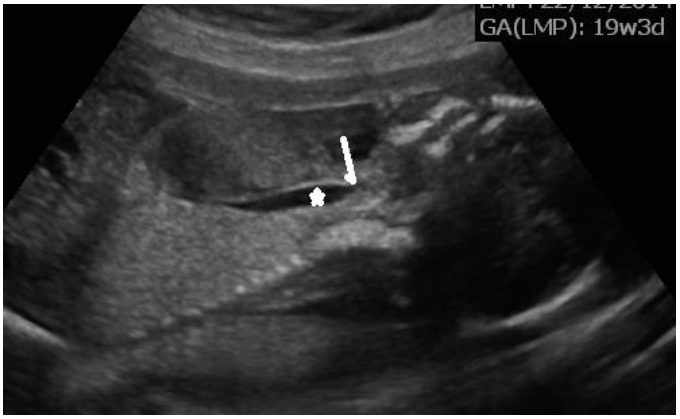


**Figure 3:** Male fetus at 19 weeks of gestation with congenital high airway obstruction syndrome. Findings: Prenatal sonographic image of fetal abdomen in axial plane showing presence of ascites (asterisk). Bold arrowhead denotes fetal liver. Technique: Clearvue 650, Philips, Convex volume transducer (2-5 MHz).



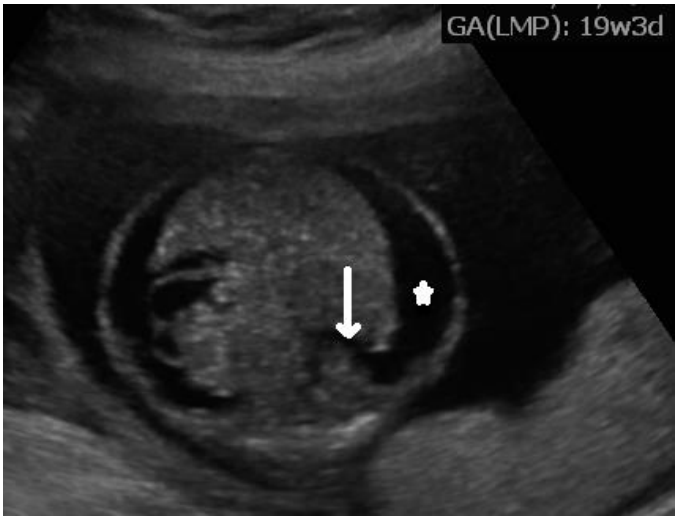




**Figure 4:** Male fetus at 19 weeks of gestation with congenital high airway obstruction syndrome. Findings: Prenatal sonographic image of fetal abdomen in axial plane showing presence of ascites (asterisk). Perinephric fluid is present (arrow), confirming hydrops fetalis. Technique: Clearvue 650, Philips, Convex volume transducer (2-5 MHz).

**Figure 5 (left):** Male fetus at 19 weeks of gestation with congenital high airway obstruction syndrome. Findings: Prenatal sonographic image of fetal thorax and cervical region in sagittal plane demonstrating dilated tracheal column (asterisk) with abrupt proximal cut off (arrow) at the level of the larynx. Technique: Clearvue 650, Philips, Convex volume transducer (2-5 MHz).

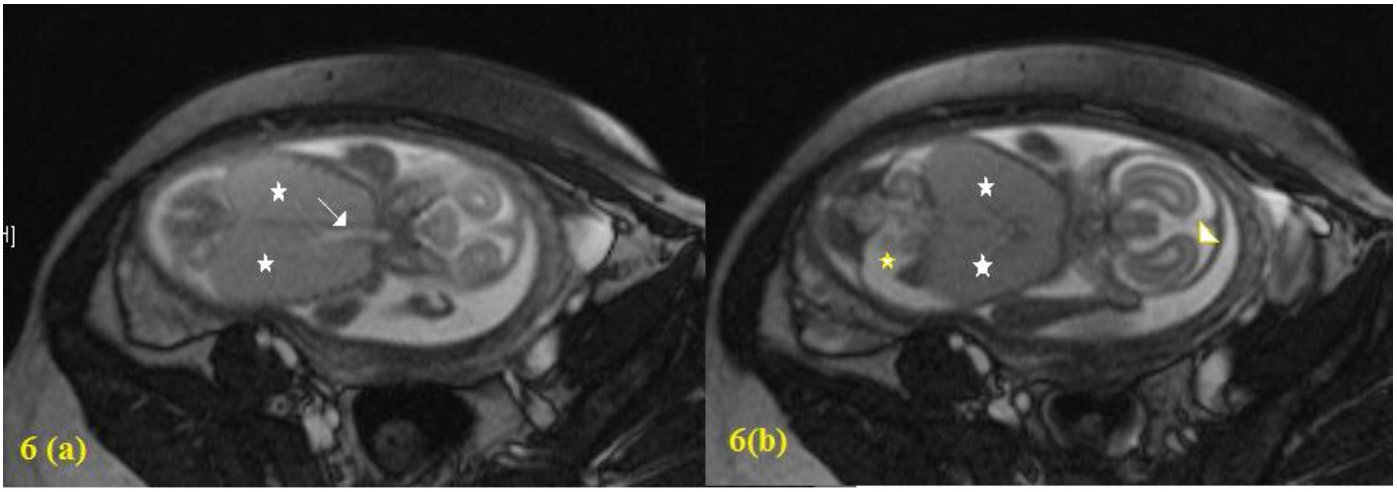



**Figure 6:** Male fetus at 19 weeks of gestation with congenital high airway obstruction syndrome. Findings: Prenatal MRI scan of fetus in coronal plane demonstrating bulky and hyperintense lungs (white asterisks) and dilated tracheal column (white arrow in Fig. 6a) confirming the prenatal diagnosis of CHAOS. Hydrops fetalis is present with ascites (yellow asterisk in Fig. 6b) and subdural collection (yellow arrowhead in Fig. 6b). Technique: 1.5T Magnet strength, Symphony, Siemens, Germany, Coronal Trufi; 346 TR; 1.4 TE, 4mm slice thickness, coronal plane.

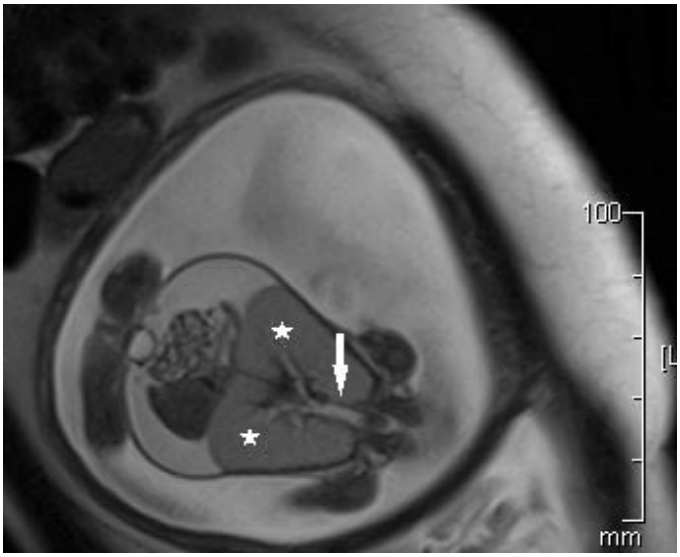



**Figure 7:** Male fetus at 19 weeks of gestation with congenital high airway obstruction syndrome. Findings: Prenatal MRI scan of fetus in coronal plane demonstrates congested lungs (asterisks) and hydrops. Arrow denotes dilated tracheobronchial tree. Technique: 1.5T Magnet strength, Symphony, Siemens, Germany, Coronal Haste 622 TR, 96 TE: 5 mm slice thickness, coronal plane.

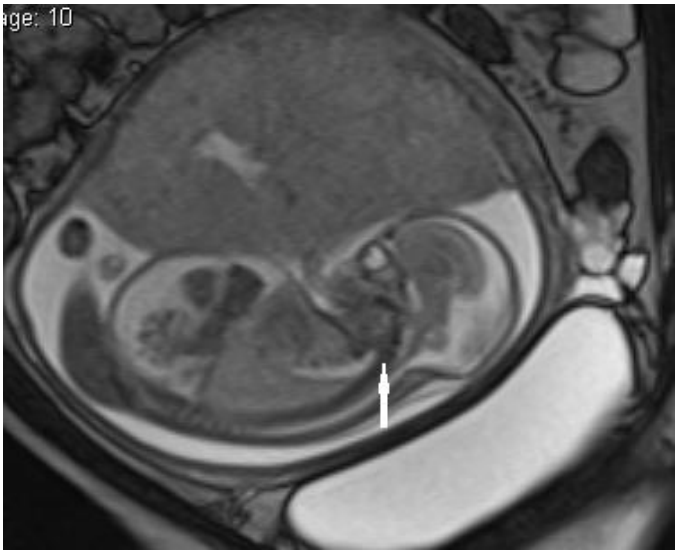


**Figure 8:** Male fetus at 19 weeks of gestation with congenital high airway obstruction syndrome. Findings: Prenatal MRI scan of fetus in sagittal plane demonstrating bulky lungs, fetal ascites and subdural collection and dilated trachea confirming the prenatal diagnosis of CHAOS. Placentomegaly is present. Tracheal dilatation with proximal cut off at the level of larynx (arrow) is demonstrated. Technique: 1.5T Magnet strength, Symphony, Siemens, Germany, Trufi; 346 TR; 1.4 TE, 3.5mm slice thickness, mid-sagittal plane.

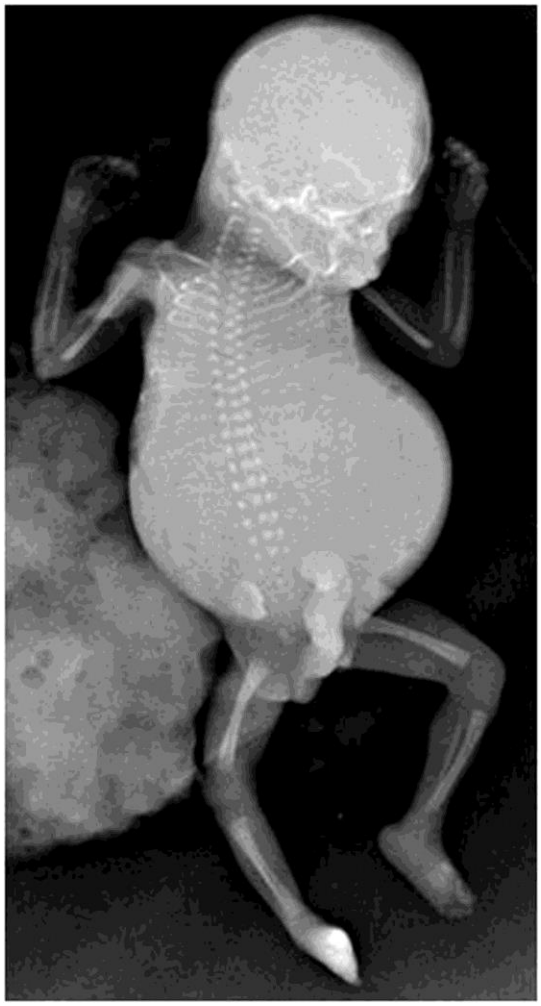
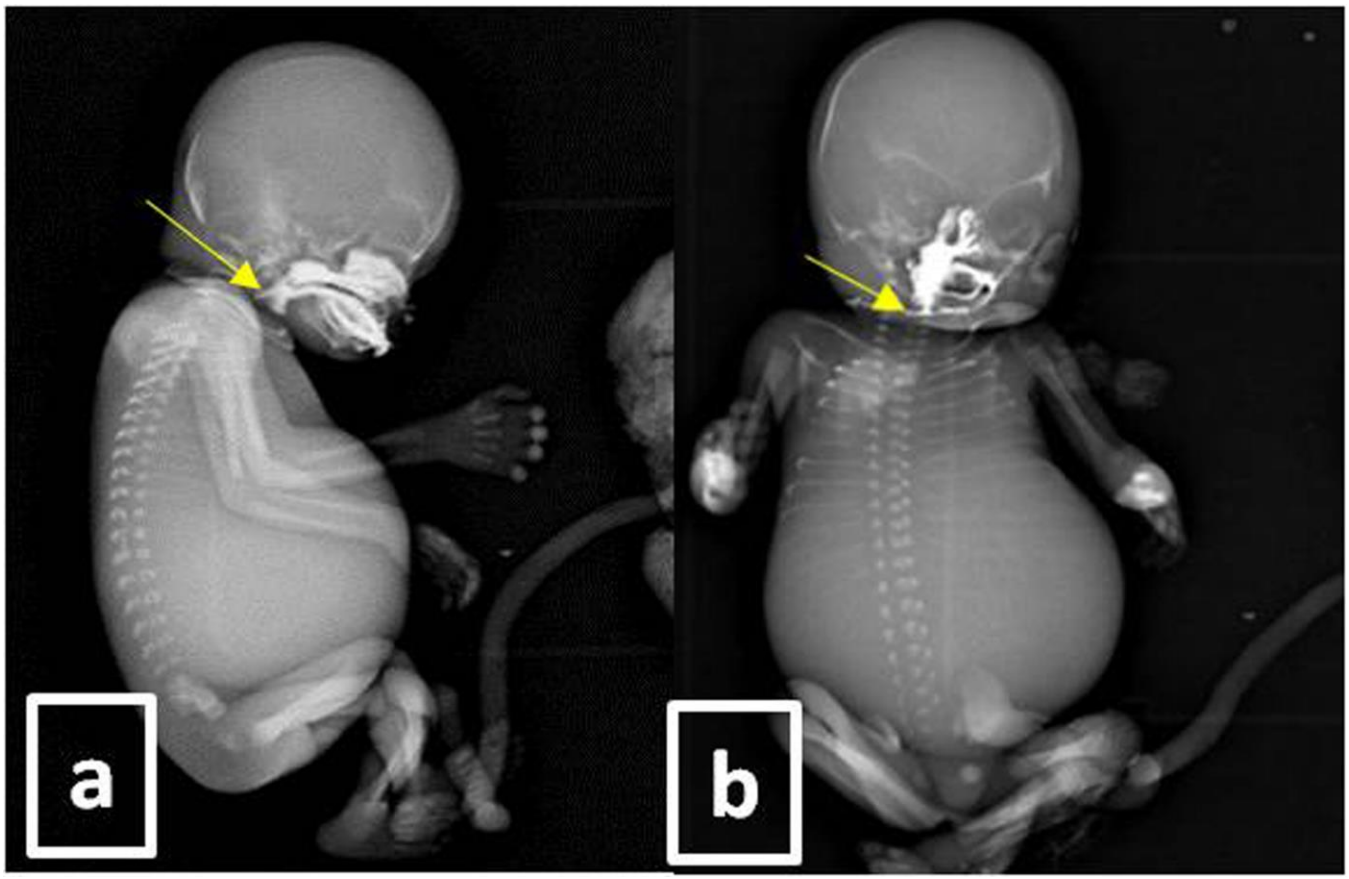


**Figure 9 (left):** Aborted male fetus at 20 weeks of gestation with congenital high airway obstruction syndrome: Findings: Plain radiograph showing gross abdominal distention and opaque thorax. Technique; KonicaMinolta Aero DR System. 53kV, 6mAS.

**Figure 10 (bottom):** Aborted male fetus at 20 weeks of gestation with congenital high airway obstruction syndrome: Findings: Contrast radiograph of upper airways in (a) lateral and (b) antero-posterior views depicting normal contrast filling of the pharynx with abrupt distal cut off (yellow arrow). Technique: Contrast (Iohexol). KonicaMinolta Aero DR System. 53kV, 6mAS.



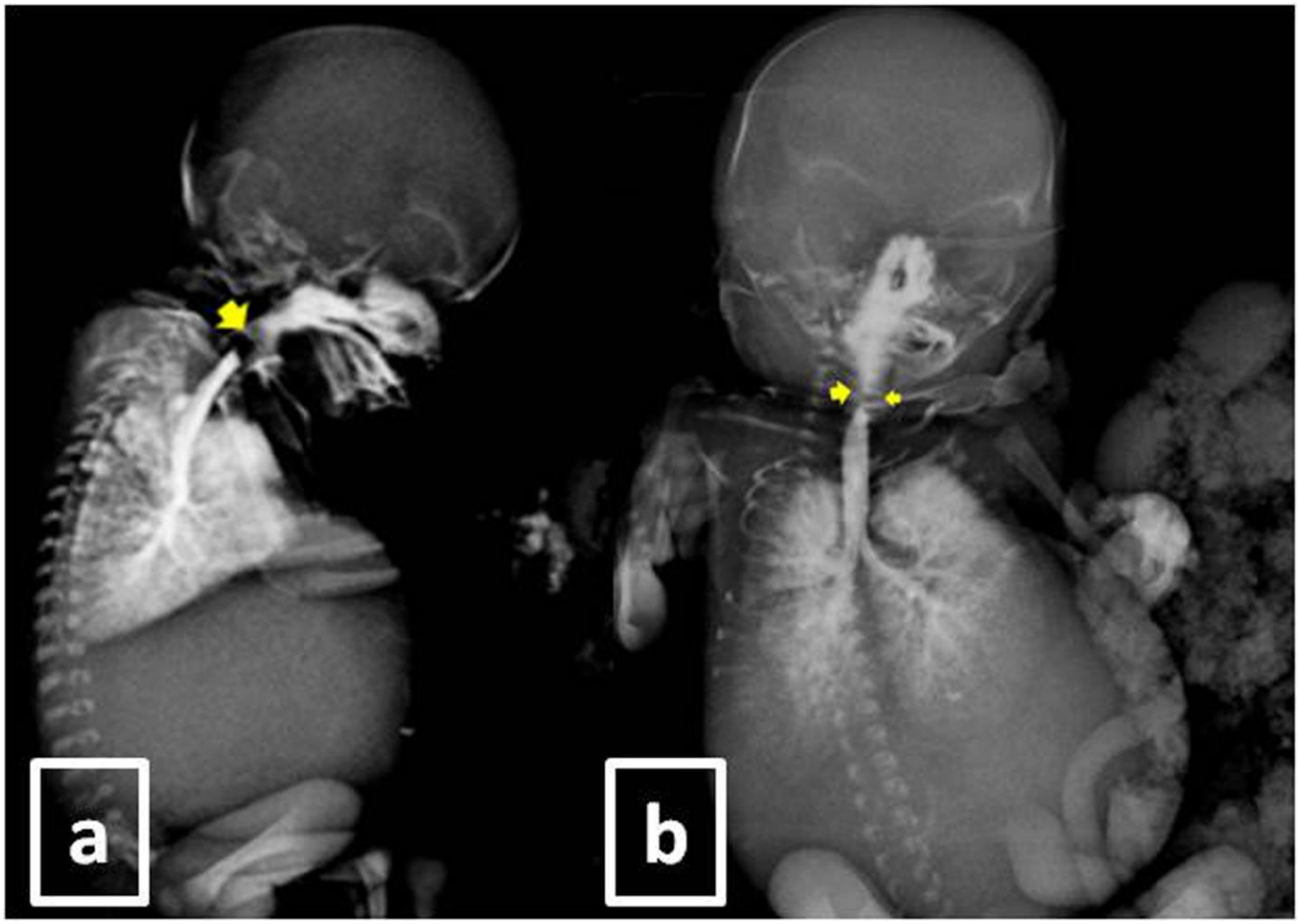



**Figure 11:** Aborted male fetus at 20 weeks of gestation with congenital high airway obstruction syndrome: Findings: Retrograde laryngogram in lateral (a) and antero-posterior (b) views demonstrating hour glass deformity (yellow arrowheads) with complete lack of contrast filling in the region of larynx. Technique: Contrast (Iohexol). KonicaMinolta Aero DR System. 53kV, 6mAS.

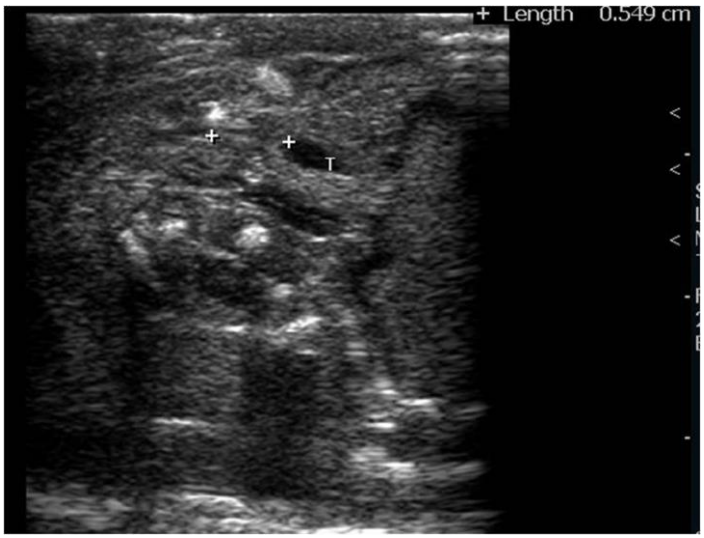



**Figure 12:** Aborted male fetus at 20 weeks of gestation with congenital high airway obstruction syndrome: Findings: Post mortem sonogram of cervico-thoracic region in sagittal plane reveals a linear column of stenosis (marked between + and +) in the region of larynx. Dilated tracheal column (T) is noted distal to stenosis. Technique: HD7, Philips, Linear transducer (3-12 MHz).

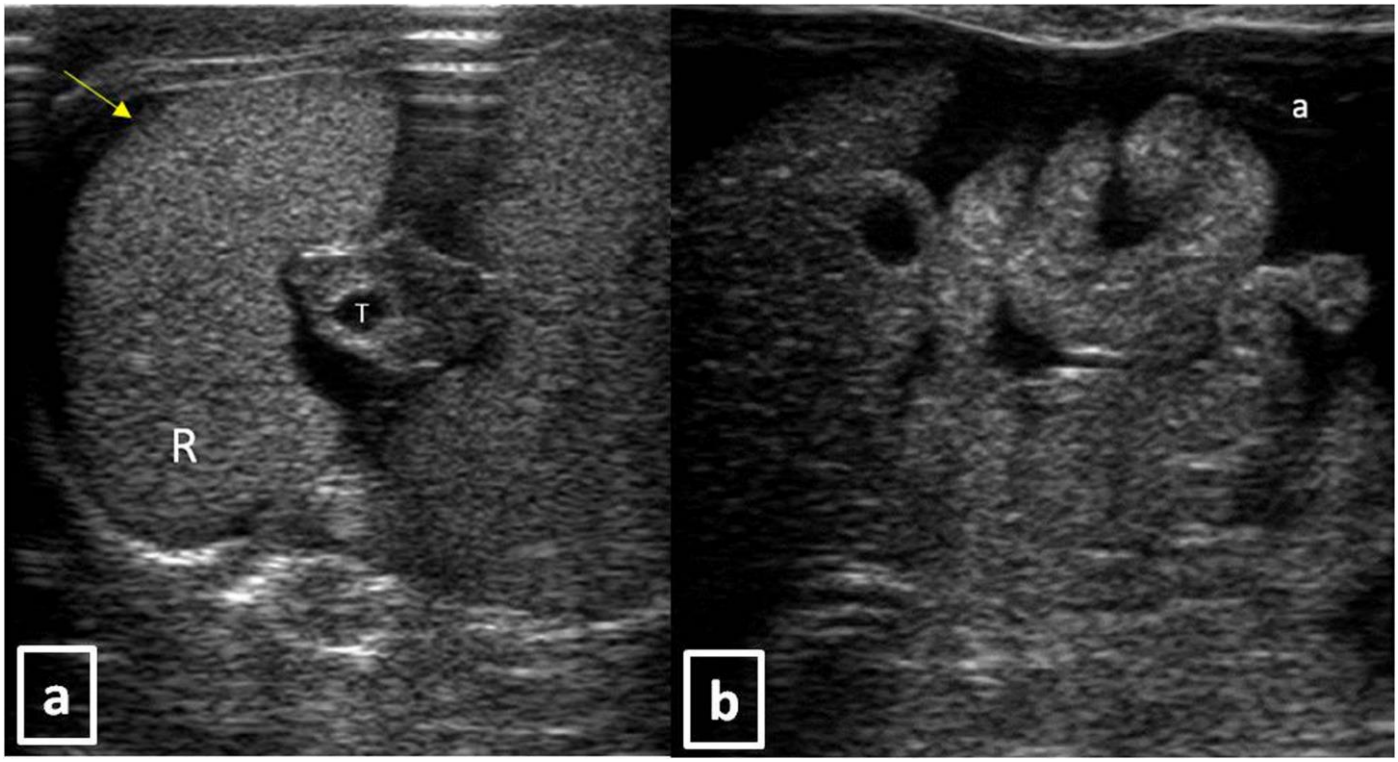



**Figure 13:** Aborted male fetus at 20 weeks of gestation with congenital high airway obstruction syndrome: Findings: Post mortem sonogram in axial plane at the level of thorax confirms echogenic and congested lungs with right pleural effusion (yellow arrow) and fetal ascites (marked by a. in Fig. 13b). Technique: HD7, Philips, Linear transducer (3-12 MHz).

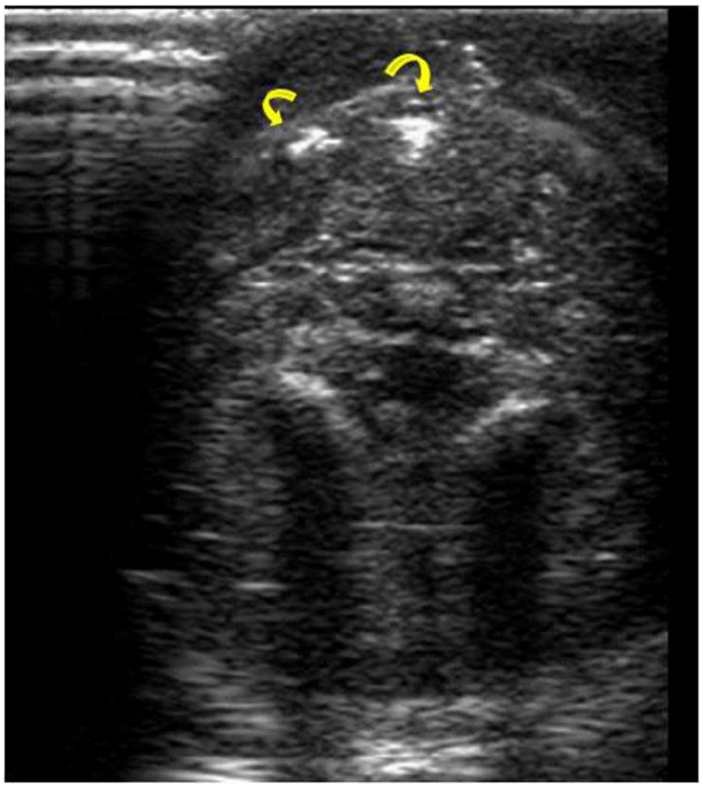


**Figure 14:** Aborted male fetus at 20 weeks of gestation with congenital high airway obstruction syndrome: Findings: Post mortem sonogram of cervico-thoracic region in axial plane reveals tracking of contrast inferiorly into blind channels (curved yellow arrows) at the level of pharynx. Technique: HD7, Philips, Linear transducer (3-12 MHz).

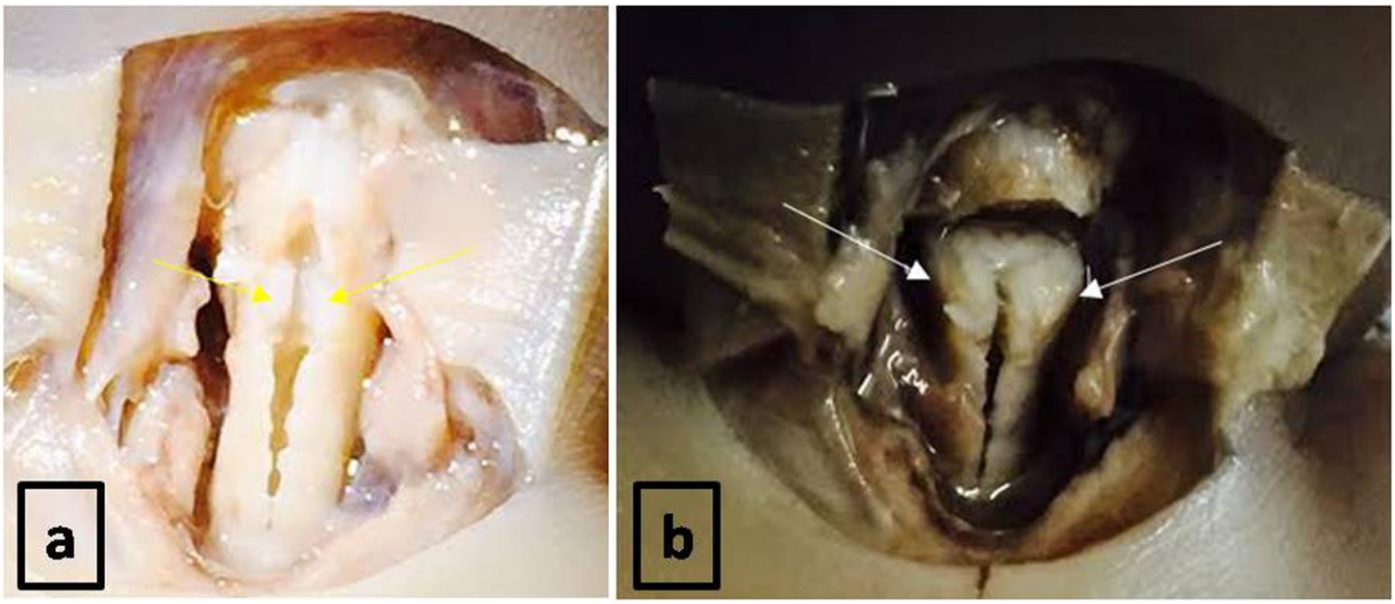



**Figure 15:** Aborted male fetus at 20 weeks of gestation with congenital high airway obstruction syndrome: Findings: Fetal autopsy revealed elliptical shaped cricoid cartilaginous ring (marked by arrows).





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|---|---|
| **Etiology** | Complete/near complete obstruction of upper airways in a fetus. |
| **Incidence** | Rare, with its true incidence unknown, 36 prenatally diagnosed cases reported till 2007. |
| **Causes** | Laryngeal or tracheal atresia, subglottic stenosis, laryngeal cyst or laryngeal web. |
| **Age predilection** | Diagnosed between 18 to 31 weeks of gestation. |
| **Gender predilection** | None. |
| **Diagnostic modality** | Ultrasound and MRI. |
| **Imaging Findings** | Bulky and echogenic lungs, inversion or flattening of diaphragm, compressed heart, dilated trachea and fetal ascites on ultrasound and MRI. |
| **Treatment** | Fetal delivery with EXIT Procedure, Fetoscopic surgery. |
| **Prognosis** | Very poor. |
| **Associated syndromes** | Most cases of CHAOS are sporadic, however known as a part of Fraser syndrome. |

**Table 1:** Summary table of key aspects and imaging findings of fetal CHAOS.

| Type of pathology | CHAOS | CCAM | Pulmonary sequestration |
|---|---|---|---|
| **Time of diagnosis** | 18 to 31 weeks of gestation. | Second and third trimester | Second and third trimester. |
| **Laterality** | Always bilateral | Bilaterality rare (2%). Usually unilobar. | Only few cases of bilaterality known. |
| **Associated early hydrops** | Always | Uncommon (5%) | Sometimes |
| **Resolution of abnormal sonographic findings** | Sometimes | Rarely | Uncommon |
| **Distinguishing imaging feature** | Dilatation of tracheobronchial tree | Circumscribed, uniformly echogenic mass supplied by a pulmonary artery. | Circumscribed, echogenic solid mass with blood supply from an anomalous systemic artery arising from thoracic/ abdominal aorta |
| **Prognosis** | 100% fatal without fetal/neonatal intervention | Perinatal mortality 9% to 49% | Good prognosis without hydrops |
| **Incidence** | Very rare with true incidence unknown | 1:11,000 and 1:35,000 live births | 0.1%. |
| **Gender Predilection** | None | Males marginally more commonly affected than females | Recognized male predilection (M:F ratio = 4/1) |

**Table 2:** Differential diagnosis table for fetal CHAOS.



## ABBREVIATIONS

CCAM: Congenital cystic adenomatoid malformation
CHAOS: Congenital high airway obstruction syndrome
EXIT: Ex-utero intrapartum treatment

## KEYWORDS

CHAOS; fetus; prenatal diagnosis; contrast radiographs; laryngeal obstruction; Retrograde laryngogram; subglottic stenosis; EXIT