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CHAOS BEFORE BIRTH: A RARE CASE OF FETAL AIRWAY OBSTRUCTION

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ABSTRACT
Congenital high airway obstruction syndrome (CHAOS) is a rare and potentially fatal condition characterized by complete or partial obstruction of the fetal upper airway, leading to a cascade of pathological changes including lung hyperexpansion, mediastinal compression, and fetal hydrops. We present a case of a 25-year-old G2P1L1 pregnant female who presented with bleeding per vaginum at 18 weeks of gestation for an antenatal anomaly scan. Ultrasound examination revealed a single live intrauterine fetus with bilateral echogenic and enlarged lungs compressing the heart, a flattened diaphragm, dilated trachea and bilateral bronchi, gross fetal ascites, hepatomegaly, echogenic collapsed bowel, and edematous abdominal skin. The nuchal fold thickness was borderline at 5.9 mm, and the abdominal circumference along with the estimated fetal weight were above the 97th percentile. These features were consistent with the diagnosis of congenital high airway obstruction syndrome, which was further supported by limited-sequence fetal magnetic resonance imaging. The patient had a history of completed treatment for pulmonary tuberculosis six months prior to conception. This case highlights the characteristic prenatal ultrasound findings of CHAOS and emphasizes the importance of early diagnosis for appropriate counseling and perinatal management planning, including consideration of ex utero intrapartum treatment procedures.

KEYWORDS: Congenital high airway obstruction syndrome, CHAOS, fetal ascites, echogenic lungs, laryngeal atresia, prenatal diagnosis

INTRODUCTION

Congenital high airway obstruction syndrome (CHAOS) is a rare and life-threatening fetal condition characterized by complete or near-complete obstruction of the fetal upper airway, which can occur at the level of the larynx, trachea, or result from extrinsic compression such as congenital neck masses (1). The true incidence of CHAOS remains unknown, though an incidence of approximately 1 per 50,000 newborns has been described in the literature (2). The underlying pathophysiological mechanism involves impaired clearance of fetal lung fluid due to airway obstruction, leading to increased intratracheal pressure and secondary hyperplastic lung growth (3). The resulting hyperexpanded, echogenic lungs compress the heart and great mediastinal vessels, causing elevated intrathoracic pressure, decreased venous return, and ultimately fetal cardiac failure, which produces the characteristic findings of fetal ascites, hydrops, flattened or inverted diaphragm, and dilated distal airways (4). Prenatal diagnosis is typically made through ultrasound examination, which remains the modality of choice for identifying the classic features of CHAOS, while magnetic resonance imaging provides superior anatomical delineation and helps identify the level of obstruction, which is crucial when planning fetal interventions (5). Without timely diagnosis and appropriate perinatal management, CHAOS carries a dismal prognosis, often resulting in stillbirth or early neonatal death; however, advances in fetal therapy, including the ex utero intrapartum treatment procedure and fetoscopic interventions, have improved survival outcomes in carefully selected cases (6). We report a case of CHAOS diagnosed prenatally at 18 weeks of gestation, illustrating the characteristic sonographic and MRI findings, and discussing the clinical implications for perinatal management.

Case Report

A 25-year-old G2P1L1 pregnant female presented with complaints of bleeding per vaginum and was referred for an antenatal anomaly scan. Her first pregnancy had resulted in an uneventful normal vaginal delivery, and there was no significant family history of congenital anomalies or genetic disorders. The patient had a history of pulmonary tuberculosis and had completed anti-tubercular treatment six months prior to conception. Her last menstrual period was dated 23 March 2026, with an estimated gestational age by last menstrual period of 17 weeks and 1 day at the time of the initial assessment, and the expected date of delivery by last menstrual period was calculated as 28 December 2026.

A transabdominal ultrasound examination was performed under good technical conditions and revealed a single live intrauterine fetus with a fetal heart rate of approximately 157 beats per minute and demonstrable fetal activity. The presentation was variable in utero, the placenta was located anteriorly, and the liquor was adequate with multiple echogenic foci, showing a single vertical deepest pocket of approximately 5.9 cm. The nuchal fold thickness measured

approximately 5.9 mm, which was considered borderline and potentially indicative of early hydrops. On thoracic evaluation, the bilateral lungs appeared markedly echogenic and enlarged, severely compressing the heart, and the trachea appeared dilated along with bilateral dilated bronchi, findings which are pathognomonic for upper airway obstruction and are illustrated in Fig. 1 and Fig. 2. The heart itself was compressed but demonstrated normal cardiac situs and a normal four-chamber view, while the diaphragm was flattened due to the downward pressure exerted by the hyperexpanded lungs (Fig. 3). Abdominal examination revealed normal abdominal situs, but the liver appeared echogenic and enlarged in size, the bowel appeared echogenic and collapsed, and there was gross fetal ascites along with edematous and thickened abdominal skin, as demonstrated in Fig. 4. The bilateral kidneys were visualized and appeared grossly normal, while the bladder appeared empty. The extremities, including all long bones, hands, and feet, appeared normal for the period of gestation. Biometric measurements showed the biparietal diameter measuring 4.05 cm corresponding to 18 weeks and 2 days (91.4th percentile), the head circumference measuring 14.74 cm corresponding to 17 weeks and 6 days (77.6th percentile), the abdominal circumference measuring 15.16 cm corresponding to 20 weeks and 3 days and falling above the 97th percentile, and the femur length measuring 2.11 cm corresponding to 16 weeks and 2 days (15.6th percentile). The estimated fetal weight using biparietal diameter, head circumference, abdominal circumference, and femur length was 239 ± 35.0 grams, which also fell above the 97th percentile, reflecting the significant organomegaly and ascites contributing to the increased abdominal girth. The overall impression was that of a single live intrauterine fetus assigned as per last menstrual period corresponding to approximately 18 weeks and 1 day, with abdominal circumference and estimated fetal weight falling above the 97th percentile, and the constellation of findings including gross fetal ascites, hepatomegaly, bilateral echogenic enlarged lungs compressing the heart, flattened diaphragm, dilated trachea and bilateral bronchi, and thickened edematous abdominal skin were highly suggestive of congenital high airway obstruction syndrome. Following the ultrasound diagnosis, limited-sequence fetal magnetic resonance imaging was performed for further evaluation and to precisely identify the level of obstruction, as shown in Fig. 5. The MRI findings were consistent with high laryngeal or high airway obstruction syndrome, demonstrating dilated bronchi, a dilated trachea, echogenic lungs, echogenic bowel, and a flattened diaphragm, which corroborated the ultrasound findings and suggested the level of obstruction to be at the laryngeal level. Following the diagnosis and extensive multidisciplinary counseling regarding the poor prognosis of CHAOS and the available management options, including the possibility of ex utero intrapartum treatment, the parents elected to terminate the pregnancy. Post-termination, X-ray of the abortus was performed, which confirmed the fetal abnormalities consistent with the prenatal imaging findings.

DISCUSSION

CHAOS represents a rare but well-recognized fetal condition that carries significant morbidity and mortality if not diagnosed prenatally, and the case presented here demonstrates the classic constellation of ultrasound findings that enable the diagnosis of this syndrome. The pathogenesis of CHAOS begins with an intrinsic or extrinsic obstruction of the fetal upper airway, and since the fetal lungs continuously produce fluid which is normally cleared through the tracheobronchial tree into the amniotic cavity, airway obstruction leads to fluid accumulation, increased intratracheal pressure, and secondary hyperplasia and hyperexpansion of the lungs (7). The massively enlarged echogenic lungs then compress the heart and great mediastinal vessels, resulting in decreased venous return, elevated central venous pressure, and ultimately fetal cardiac failure, which produces the characteristic findings of ascites, hydrops, and flattened or inverted diaphragm (8). In the present case, all these features were observed: bilateral echogenic enlarged lungs compressing the heart (Fig. 1), flattened diaphragm (Fig. 3), gross fetal ascites with edematous abdominal skin (Fig. 4), and the presence of hepatomegaly and echogenic collapsed bowel further supported the diagnosis, likely reflecting the hemodynamic compromise and venous congestion resulting from the elevated intrathoracic pressure. The borderline nuchal fold thickness of 5.9 mm observed in this case may reflect the early stages of hydrops fetalis, as lymphatic drainage is compromised due to the elevated central venous pressure. In this case, the patient had previously normal clinical reports and an uncomplicated first pregnancy but subsequently presented with acute symptoms, highlighting the unusual and distinctive nature of this case.

Ultrasound remains the primary modality for prenatal diagnosis of CHAOS, and the condition can be diagnosed as early as 15 weeks of gestation (2). The characteristic sonographic features include large echogenic lungs that appear hyperechoic and enlarged often occupying most of the thoracic cavity, mediastinal compression with the heart displaced centrally due to the expanded lungs, a flattened or inverted diaphragm due to downward pressure, dilated fluid-filled trachea and bronchi distal to the obstruction, and fetal ascites or hydrops due to impaired venous return and cardiac failure (9). In our case, the ultrasound findings were classic: bilateral echogenic enlarged lungs (Fig. 1), compressed heart, flattened diaphragm (Fig. 3), dilated trachea and bronchi (Fig. 2), gross ascites with edematous skin (Fig. 4), and the polyhydramnios was noted with multiple echogenic foci in the liquor. Fetal MRI provides additional diagnostic information and is particularly valuable in identifying the exact level and nature of the airway obstruction, demonstrating large lung volumes, increased lung signal intensity, inverted diaphragm, and dilated fluid-filled lower airway (5). In a study by Mong et al., MRI identified the obstruction level in 90% of cases and changed the screening ultrasound diagnosis in 70% of cases, highlighting the complementary role of MRI in the diagnostic workup of CHAOS (1). In the present case, limited-sequence MRI confirmed the ultrasound findings and suggested the level of obstruction to be at the laryngeal level (Fig. 5), which is the most common site of obstruction in CHAOS.

The differential diagnosis of CHAOS includes other causes of fetal echogenic lungs and hydrops, such as congenital cystic adenomatoid malformation type III, pulmonary sequestration, congenital diaphragmatic hernia, and nonimmune hydrops fetalis from other causes (10). The presence of dilated airways distal to the obstruction is a key differentiating feature of CHAOS, as it indicates the site of obstruction, and in our case, the dilated trachea and bronchi along with the other

characteristic findings helped establish the diagnosis with confidence. CHAOS is a life-threatening condition with a historically poor prognosis, and without intervention, the condition is almost uniformly fatal, resulting in stillbirth or early neonatal death (11). However, advances in perinatal management have improved outcomes in selected cases, and the ex utero intrapartum treatment procedure, which involves partial delivery of the fetus while maintaining uteroplacental circulation to secure the airway before clamping the umbilical cord, has significantly improved survival rates (6). In a comprehensive review by Roybal et al., 36 out of 118 patients (30.5%) were alive at the time of reporting following ex utero intrapartum treatment procedures, and fetoscopic interventions including fetoscopic laser laryngotomy and tracheal decompression have also been attempted with variable success (6). The prognosis is influenced by several factors including the presence of associated anomalies, the level and completeness of obstruction, and the development of hydrops, and isolated CHAOS without other major anomalies generally carries a better prognosis than cases with associated anomalies (11). In our case, the presence of gross ascites, hepatomegaly, and edematous skin suggested established hydrops, which portends a poorer prognosis, and after extensive counseling, the parents elected for termination of pregnancy.

This case underscores the importance of meticulous prenatal ultrasound screening for early detection of CHAOS, and the characteristic findings once recognized should prompt further evaluation with fetal MRI to confirm the diagnosis and identify the level of obstruction (Fig. 5). Early diagnosis is crucial for parental counseling, as parents should be informed about the condition, its prognosis, and available management options, and for perinatal planning, multidisciplinary involvement of maternal-fetal medicine specialists, neonatologists, pediatric surgeons, and anesthesiologists is essential for optimal management (12). In cases where continuation of pregnancy is chosen, delivery should be planned at a tertiary center with ex utero intrapartum treatment capability. The patient in this case had a history of treated pulmonary tuberculosis, and while there is no established association between maternal tuberculosis and CHAOS, this history was noted and did not appear to be etiologically related to the fetal condition.

CONCLUSION

Congenital high airway obstruction syndrome is a rare but serious fetal condition characterized by complete or partial obstruction of the upper airway, leading to a cascade of pathological changes including lung hyperexpansion, mediastinal compression, and fetal hydrops. The diagnosis is primarily made through prenatal ultrasound, which reveals the classic features of bilateral echogenic enlarged lungs, mediastinal compression, flattened diaphragm, dilated distal airways, and fetal ascites or hydrops. Fetal MRI provides additional information regarding the level and nature of obstruction and is invaluable for surgical planning. Early prenatal diagnosis is essential for appropriate parental counseling, perinatal planning, and consideration of fetal interventions such as the ex utero intrapartum treatment procedure. This case report contributes to the growing body of literature on CHAOS and highlights the importance of recognizing its characteristic sonographic features for timely diagnosis and management, ultimately aiming to improve perinatal outcomes in these complex cases.

Figures

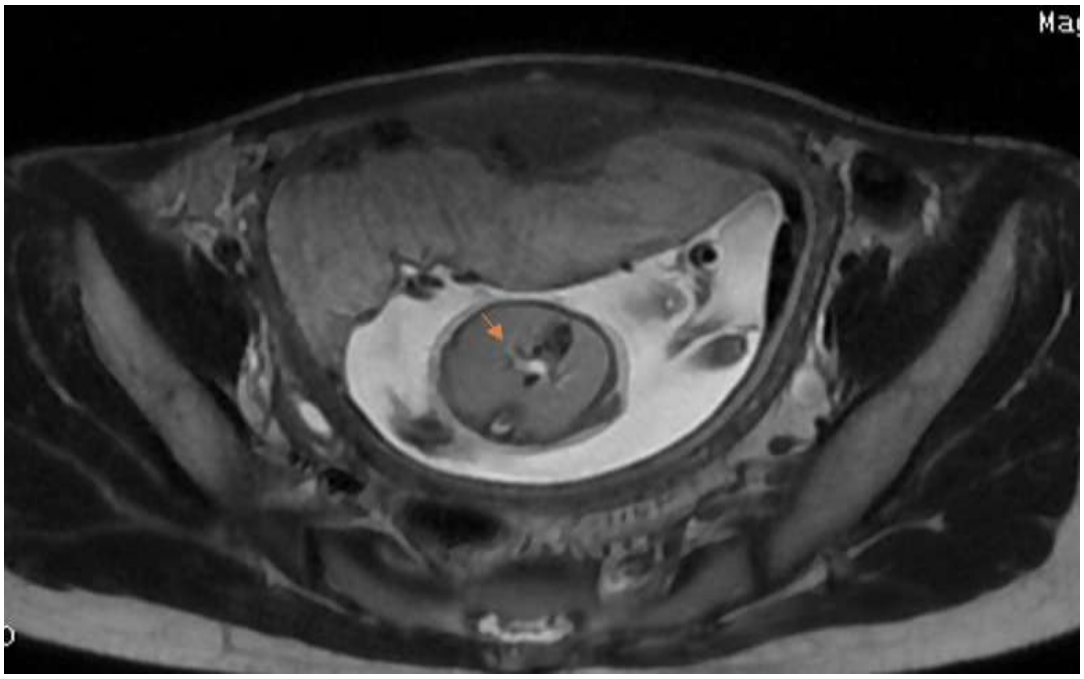

Fig. 1: Ultrasound image showing dilated bilateral bronchi, indicative of fluid accumulation and increased pressure distal to the site of upper airway obstruction, a classic finding in CHAOS.

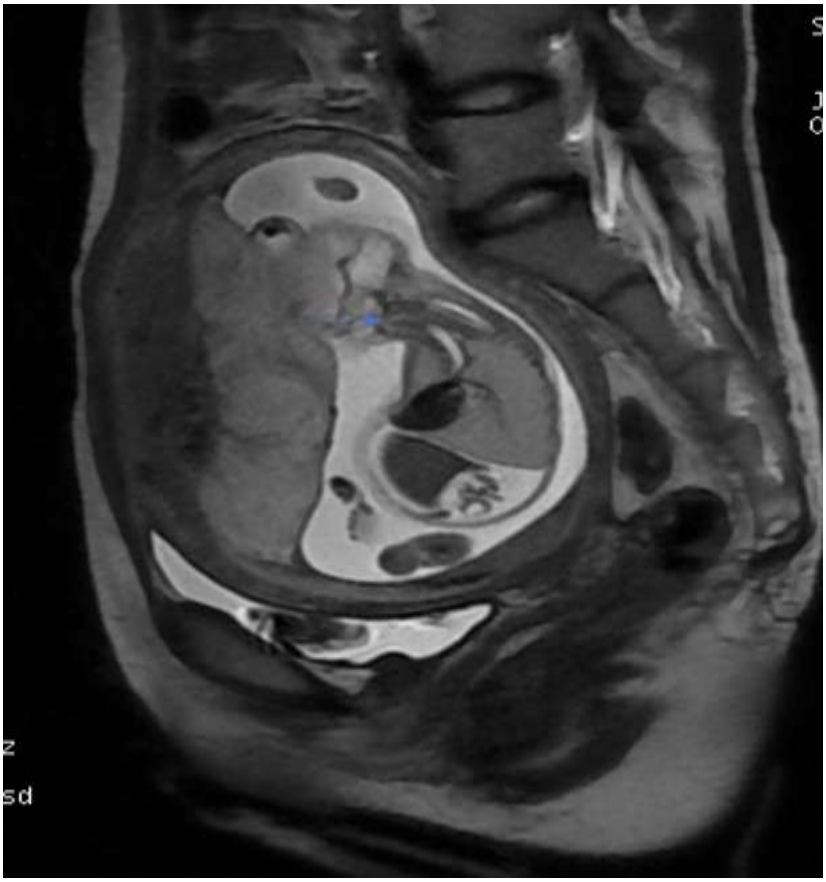

Fig. 2: Sagittal ultrasound view of the fetal thorax showing the dilated trachea and bilateral dilated bronchi filled with fluid, reflecting the accumulation of pulmonary secretions distal to the site of high airway obstruction, a hallmark feature of CHAOS.

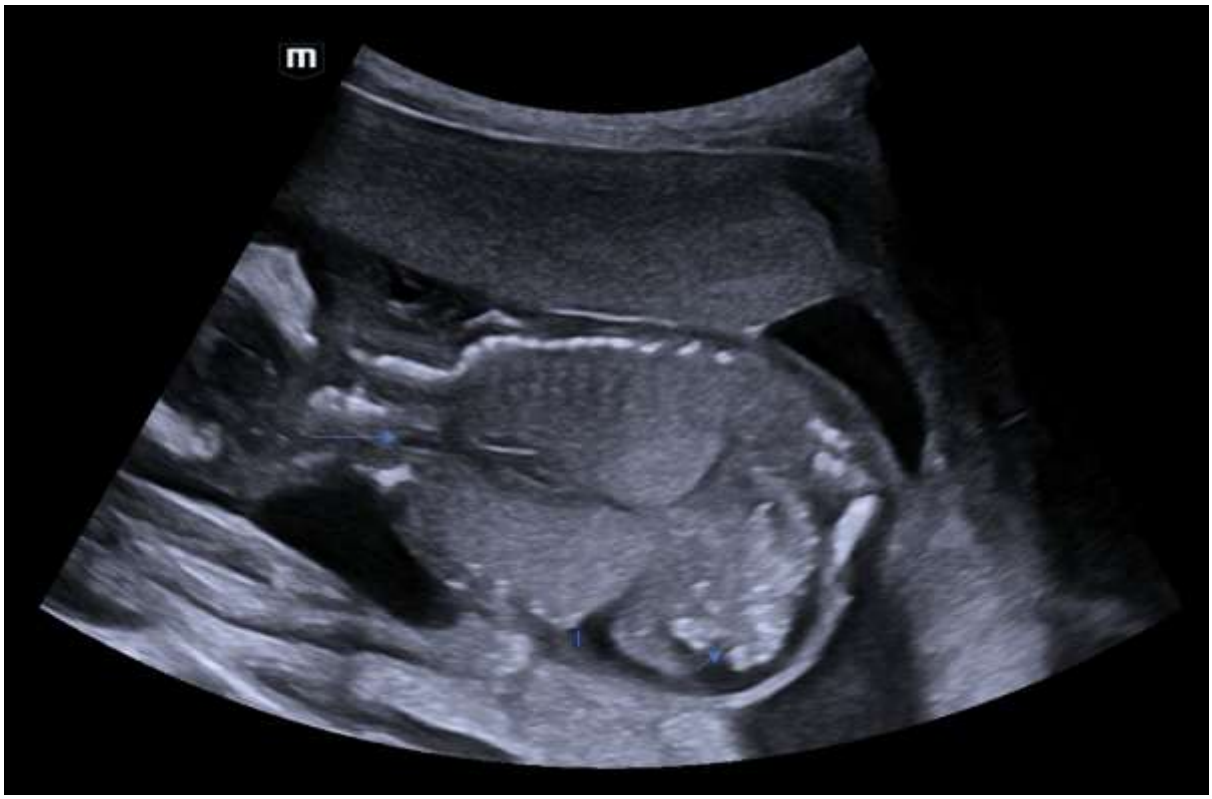

Fig. 3: Ultrasound image demonstrating a dilated trachea, hyperechogenic lung parenchyma, echogenic collapsed bowel, and a flattened diaphragm due to the downward pressure exerted by the hyperexpanded lungs.

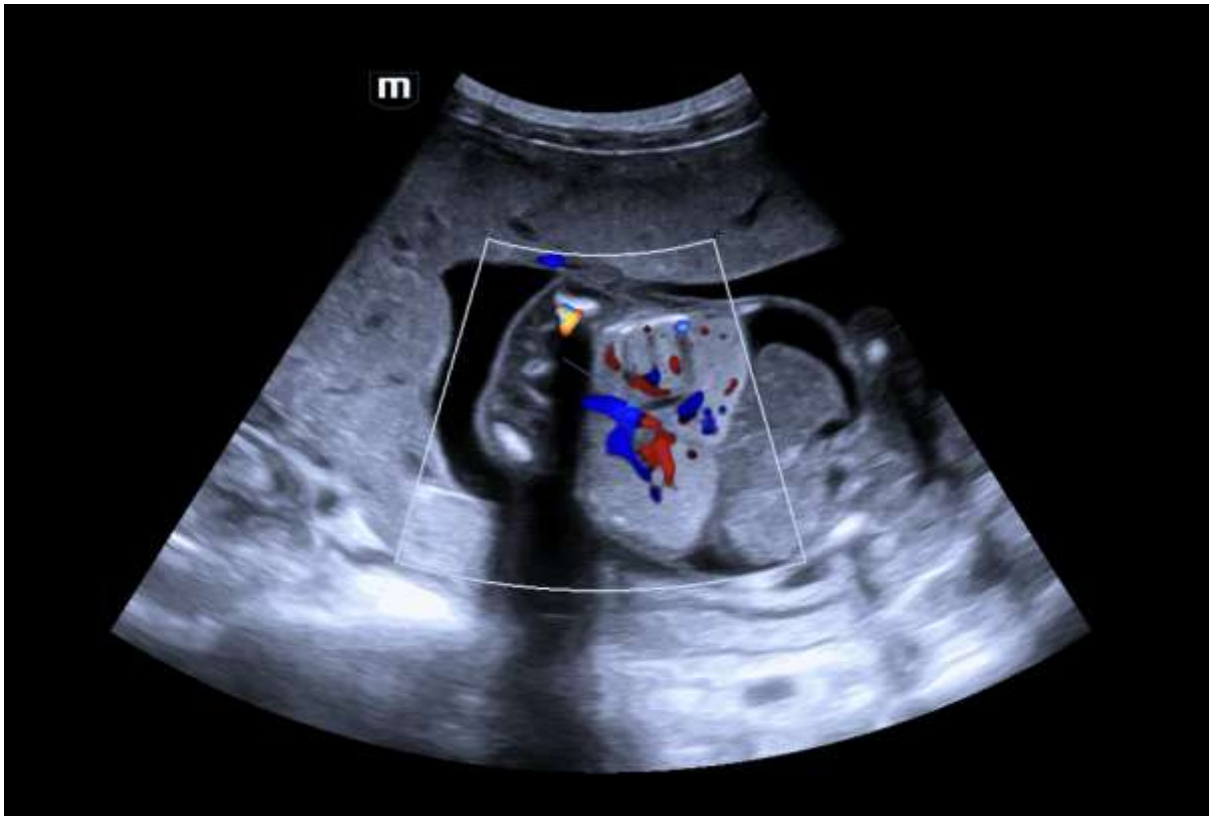

Fig. 4: Abdominal ultrasound image demonstrating gross fetal ascites with a large anechoic fluid collection surrounding the abdominal viscera, along with echogenic and enlarged liver (hepatomegaly), echogenic collapsed bowel, and markedly thickened and edematous abdominal skin, reflecting the hemodynamic compromise and cardiac failure secondary to the airway obstruction.

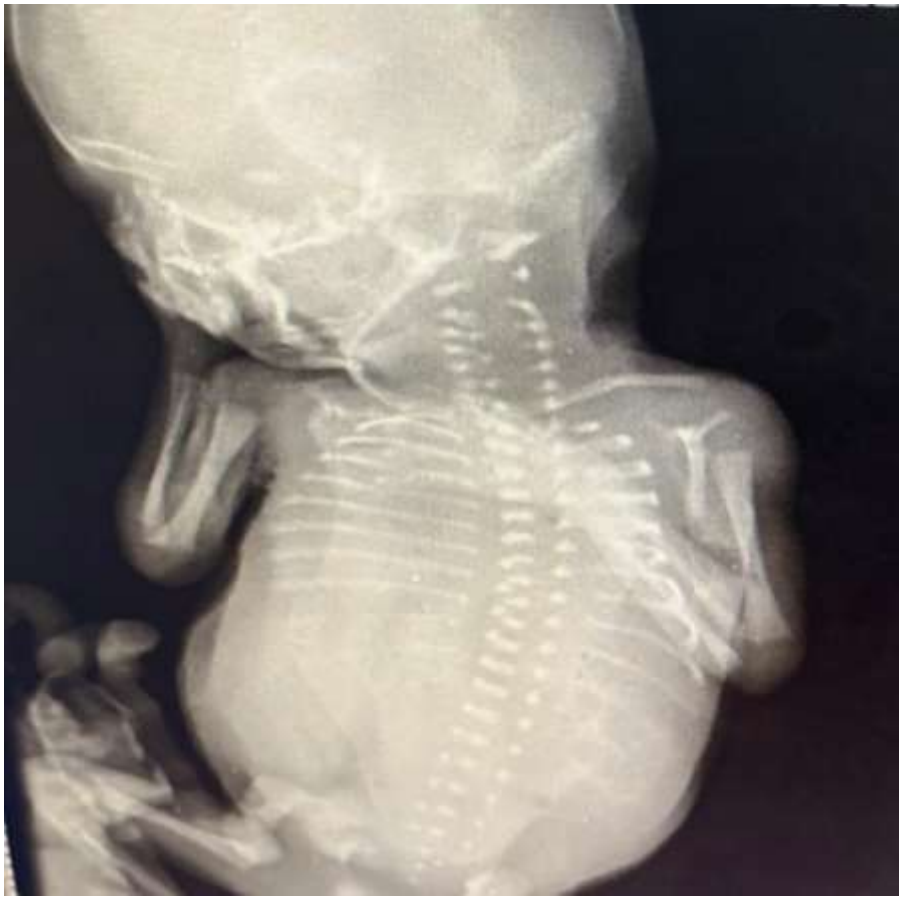

Fig. 5: Post-termination X-ray of the abortus fetus confirming the fetal abnormalities, including thoracic distension and ascites, consistent with the antenatal diagnosis of congenital high airway obstruction syndrome.

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