Congenital unilateral hyperlucent lung in pediatrics, finding in images of five cases.

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SUMMARY
The unilateral hyperlucent lung is part of the congenital anomalies of the foregut primitive, represent a broad spectrum of congenital anomalies, which have a common origin at a particular point of embryological development foregut / lung. (1) Usually they detected in the neonatal period or early childhood, although asymptomatic cases may be diagnosed in adulthood. Currently most are diagnosed in routine prenatal ultrasounds. (1)

It is a rare entity in children presenting with acute respiratory distress progressive. The chest radiograph is the primary imaging modality in the diagnosis, they are characterized by increased unilateral hyperlucent lung with displacement of the structures of the mediastinum to the contralateral side and could become evident small thin - walled air cavities.

RESUMEN
El pulmón hiperlucido unilateral forman parte de las Anomalías Congénitas del Intestino Primitivo Anterior, representan un amplio espectro de anomalías congénitas, que tienen un origen común en un punto concreto del desarrollo embriológico del intestino anterior/pulmón. (1)

Suelen detectarse en el periodo neonatal o infancia temprana, aunque los casos asintomáticos pueden diagnosticarse en la edad adulta. Actualmente la mayoría, se diagnostican en las ecografías prenatales de rutina. (1)

Es una entidad poco frecuente en pediatría que cursan con dificultad respiratoria aguda progresiva. La radiografía del tórax es la principal modalidad de imagen en el diagnóstico, se caracterizan por el incremento de la radiolucidez pulmonar unilateral con desplazamiento de las estructuras del mediastino hacia el lado contralateral y podrían evidenciarse pequeñas cavidades aéreas de paredes delgadas.

Keywords: Cystic adenomatoid malformation (MAQ). Congenital Lobar Emphysema (ELC) Diaphragmatic Hernia (HD) and Pulmonary Cysts (QP).

TARGET
Present radiology clinical cases that are part of hyperlucent lung and its main differential diagnoses, to be rare pathology is important to know to suspect and an early diagnosis, since the delay may be associated with increased morbidity and mortality. Pathogenesis, diagnosis, and prognosis of this disease is discussed.

Case No. 1:
Male patient 1 month 19 days old, goes to hospital with cough and breathlessness of 24 hours of evolution, oxygen Hood administered by show saturation 78%, FC 151 x min, tachypnea FR 100xmin, T: 37.5 C , the physical examination: grunting, malaise, subcostal and intercostal retractions and not left vesicular murmur is heard. is performed CRX and interpreted as left pneumothorax (Figure 1A), and chest tube is placed without presenting clinical improvement so it is transferred to the nursing home presenting distress frank respiritory, bad input bilateral air is not auscultates vesicular left murmur, desaturation and a thorax expanded by barrel, standard chest radiography (figure 1 B) is repeated and CT of the chest (figure 1 C), which establishes the diagnosis of cystic adenomatoid malformation (MAQ) and the patient is requested is submitted He left lobectomy.

Figure 1:
A: Rx AP chest: Signs of air trapping in the left lung field with large mediastinal shift to the contralateral side, observing the presence of vascular pattern in this hemithorax with which rules out the possibility of pneumothorax.
B: Rx AP chest: subcutaneous emphysema and chest tube is shown in the left lung, mediastinal deviation persists.
C: TC chest; multiple thin walled cystic spaces, greater than 2 cm distributed in the left lung field right accompanied by mediastinal deviation, marginal pneumothorax and subcutaneous emphysema. Findings regarding MAQ.

Figure 2: Macroscopic part of the left lung:
Histopathological results:
Macroscopic: lung lobe weighing 50.5g, measures 10 x 6 x 2cm fluffy. Injury microscopic pulmonary parenchyma with distortion of the architecture by the presence of cystic structures by ciliated and cuboidal coated columnar epithelium. These cysts communicate with each other and structures coated honeycomb cuboidal epithelium is identified.
Conclusion: Cystic adenomatoid malformation lung tipe 1.

Case No. 2:
Male patient, 18 days old, a product of the second pregnancy, regular prenatal visits, delivery by caesarean section is performed with APGAR at one minute and five minutes of 8 and 10 respectively, has a birth weight 3000gr.
Consultation breathlessness 3 days of evolution, accompanied by cough, nasal flaring, intense intercostal retractions, tachypnea and cyanosis, rales and crackles are heard in the left lung field so it is brought to this nursing home where she performs Rx chest (figure 3A) and CT chest (figure 3 B and C) Mal suspected adenomatidea cystic formation (MAQ) and the patient undergoing lobectomy is left.

Figure 3:

A: Rx AP chest: Signs of air trapping with cystic radiolucent images of thin walls accompanied by mediastinal shift at the left lung capo.

B and C: chest CT: S and confirms the presence of multiple air cavities finidas walls, diffusely distributed, with resizable less than 2 cm in relation to MAQ

Cystic adenomatoid malformation (MAQ).

Topic Review
Cystic adenomatoid malformation (MAQ) is defined as an abnormal proliferation of lung tissue secondary to an error in the maturation of bronchiolar structures. First described by Bartholin (1697), and subsequently by Ch'in and Tang (1949) who define it as a congenital malformation of the lung probably caused by an embryological aggression occurred between 40 to 50 days of gestation, and consisting of a lack of fusion of the bronchial and alveolar buds mesenchyme, leading to an abnormal and excessive development, primarily from terminal bronchioles. Its origin is due to a failure in the maturation of bronchial structures (between weeks 7-17) in which the lung tissue adopts a cystic-adenomatous appearance. (2)

It is known as a congenital malformation of the airway, is a rare entity which constitutes 25% of all lung congenital anomalies, unproved a higher incidence by sex or race. In a Sept. 5% of cases affecting a lobe of a lung, bilateral involvement is rare. (3)

Frequency
Its frequency is low, from 1 per 5,000 to 1 per 20,000 births. It is a rare congenital lung disorder, between 80-95% of cases are detected in the neonatal period and approximately 20% after this period. (2)

Characteristics
Morphologically it has characteristics of lung immaturity with cystic dilatation of the bronchioles. The MAQ communicates with the airway. The blood supply is usually done
through the pulmonary arteries, although there have been reports of systemic arterial supply (in the hybrid-type pulmonary sequestration MAQ). The venous drainage usually depends on the pulmonary veins. (3)

Expansive lesions lead to progressive symptoms. If they are large they can compromise lung development in the fetal period, appearing hypoplastic lungs. Mass effect may prevent adequate cardiac contractility and compression of the vena cava with fetal hydrops appearance. If you compress the esophagus appears Polyhydramnios to prevent swallowing amniotic fluid. (3)

In children and adults may be asymptomatic, appearing as pneumonic boxes repeat most common clinical manifestation when they are symptomatic. Have been described repeated pneumothorax, obstructive bronchitis and chest pain, but are less frequent. (3)

Classification
Since 1978 it has a pathological classification given by Stocker, based on size, which derive their clinical and prognostic implications:
Type I: few large cysts that are greater than 2 cm and may reach 10 cm (50 to 65% of cases). Is the variety of better prognosis.
Type II: undersize numerous cysts (0.5 to 2 cm), uniform associated with other malformations, among which one can find normal alveoli (25% of cases). The prognosis depends on the existence of associated injuries.
Type III: rarer, solid mass formed by multiple millimetric cysts (<0.5 cm).

His association with other malformations is rare (10%). Worse prognosis. (2)
Since 1999, the author divides into five types, which are also used cri histopathologic rivers. Its diagnosis can be made prenatally by obstetrical ultrasound, or be postnatal through radiological and pathological techniques. (2)

Clinic
Usually diagnosed prenatally, during the neonatal period or during the first two years of life, being strange its late diagnosis (only 17% of cases). (3) Clinically it is associated with three common forms of presentation: 1) Perinatal death, 2) progressive respiratory distress syndrome, 3) late diagnosis (2)
On clinical examination can be found decreased breath sounds, heart sounds distant, hyperresonance side affection and prominence of the ipsilateral hemithorax. (3)

Imaging findings.
This may be suspected prenatally entity (between 18 and 24 weeks) by ultrasound when a solid mass or cystic identified in one of the fetal lungs. As secondary to the mass effect of MAQ associated findings may be mediastinal shift to the contralateral hemithorax, polyhydramnios and fetal hydrops. (3)

In the neonatal period and childhood may be performed thoracic ultrasound to diagnose this entity, showing a dominant cyst surrounded by smaller cysts in the case of MAQ type I cysts of uniform size in the case of MAQ type II and a mass solid in the case of type III MAQ. (3)

Similarly to what happens in ultrasound, simple and CT radiography the MAQ manifest as unilateral lesions with multiple rounded and thin - walled air cysts, varying the number and size of cysts according to the type of MAQ, expansion corresponding hemithorax and
mediastinal shift. The most common clinical manifestation are pneumatic repetition boxes that appear as fluid levels in image inside cysts or solid appearance thereof.\(^{(3)}\)

**Treatment and prognosis.**
In symptomatic individuals treatment of choice is surgical removal of the lobe with or sits segment where the lesion, which can be performed if necessary utero. If conservative treatment of asymptomatic postulated although there are authors who support the surgical treatment in these patients because of the risk of recurrent infections and, although questionable, the risk of malignancy.\(^{(3)}\)

**Case No. 3:**
RN term delivery product vaginal cephalo who from birth has difficulty breathing. Physical examination shows subcostal retractions and absence of vesicular murmur predominantly left.

![Image](image.png)

Figure 4:
A: Rx: left lung hyperinflation is accompanied by pulmonary herniation and diversion structures of the mediastinum to the contralateral side, suggestive of congenital lobar emphysema.

### Congenital lobar emphysema (ELC).

**Incidence**
The ELC is from 1.4 to 2.2\% of all congenital malformations. The incidence is 1 case per 20,000 to 30,000 live births and is predominant in males (3: 1).\(^{(7)}\)

**Topic Review**
Bronchi allow entry of air inspired during exhalation but with subsequent collapse of hyperinflation one or more lobes in histologically normal lungs. This overinflating causes a focal air retention to expiration with the consequent relaxation of the affected lobe and compression of adjacent structures. According to the theory suggested by Hislop and Reid defect is based in poor cartilage development (bronchomalacia) that sustains the bronchus of the affected lobe or in an intraluminal obstruction. In half of the cases no cause is found\(^{(7)}\).

It usually involves a single lobe; in order of frequency, the most affected is the left upper lobe (42.2\%), followed by the right middle lobe (35.3\%) and the right upper lobe
The lower lobes are often not involved (0.9%). Multilobar injury, as well as a bilateral lungs and are extremely rare forms. (8)

Although not rare, its frequency is difficult to pinpoint. They are responsible for significant morbidity in newborns, infants, children and even in adults, and represent the second leading cause of premature mortality in infants after abnormalities of the cardiovascular system. (8)

The ELC is associated with other congenital malformations in 14-40% of cases, the most common being the ductus patent ductus arteriosus, interventricular septal defects, diaphragmatic hernia and kidney malformations. (7)

Macroscopic lung lobe view shows a spongy appearance and yellowing, no volume decreases with compression, and emphysematous microscopically large areas are seen within the affected lobe, without destruction of the alveolar walls. (8)

Clinic
It is a disease whose clinical presentation mainly occurs in early childhood, although cases have been reported in which there was no symptoms until after 5-6 months of life, and even asymptomatic cases. (8)

The clinic usually begins in the neonatal period, usually with tachypnea, dyspnea, coughing, cyanosis, wheezing, decreased breath sounds on the affected side of the chest and crackles, symptoms obey the lobar hyperinflation and bronchial obstruction. In patients in the clinic begins around the year it may appear as recurrent respiratory infections. Although the diagnosis may be suspected on clinical examination, must be confirmed by imaging tests. (8)

Classification
It usually manifests in the neonatal period or early childhood with respiratory distress. Myers described three clinical types depending on whether congenital lobar emphysema becomes evident in early childhood (type I), in older children (type II) or as an incidental finding in asymptomatic patients (type III). Types II and III are rare. (5)

Imaging studies
Congenital lobar emphysema is a clinicopathological entity characterized by over-distension and air trapping in the affected lobe, concomitant compression of the remaining lung tissue and mediastinal shift by herniation of emphysematous lobe to the contralateral lung. (8)

The ELC is one of the causes of unilateral lung hiperlúcido, radiographs (AP) and lateral chest are essential in the diagnosis and monitoring of these entities. In the chest radiograph we can observe a strain on affected lobe or lobes with poorly defined lines inside bronchovascular, contralateral lung atelectasis, flattening of the ipsilateral hemidiaphragm and mediastinal shift to the contralateral side. Computed tomography (CT) of the chest demonstrates the high resolution and compression zone emphysematous parenchyma unaffected. (7)

The characteristic radiologic findings progressive affected lobe hyperinflation with air trapping during expiration and compression of the adjacent lobe or lobes. There bronchovascular presence of marks on the lobe on distended, useful to differentiate it from
a pneumothorax or pulmonary cyst finding. If the affected lobe is markedly expanded is possible mediastinal shift, separation of ipsilateral hemidiaphragm ribs and depression. (Figure 2) 

**Treatment**

Usually considered surgery (lobectomy) as the first treatment option in symptomatic patients (6)

Lobectomy has been the traditional treatment in patients with ELC having severe symptoms or onset neonatal stage. Today, conservative treatment is the most acceptable in asymptomatic children of any age or who have mild or moderate respiratory symptoms, especially if the bronchoscopic examination is normal. In these cases the conduct to follow will close observation and monitoring, as many cases regress. Conservative management in children can be carried out under strict monitoring, providing information about the disease and its complications parents and training them to identify the alarm data. In case of persistent respiratory symptoms lobectomy should be performed electively, thus avoiding higher risks for the patient. (7)

**Case No. 4:**

Figure 5: 
A: representative of diaphragmatic hernia Scheme. 
B and C: Rx AP and side: Left posterior diaphragmatic hernia (Bordalek) is evident. Presence of stomach and bowel loops in the left chest that displace the mediastinum to the contralateral side.

**CDH (HDC)**

It is an anatomical defect of the diaphragm of unknown etiology that is associated with significant morbidity and mortality. (14)

**Incidence:**
It is about 1 per 2 200 2 400 live births, with a mortality rate ranging from 40 to 80%. (14)

**Topic Review:**
The HDC is a result of the failure to close the pleuroperitoneal duct, around 9 to 10 weeks gestation, at which time midgut returns from the yolk sac into a relatively small peritoneal cavity, which favors its herniation through diaphragmatic defect, accompanied by other organs such as stomach, liver or spleen. (14)
The fetal lung development is controlled primarily by mechanical (strain or stretching) forces, whose molecular effectors are growth factors, especially those derived fibroblasts
(FGF). The herniated mass interferes with these forces and induces variations in the molecular signals that result in pulmonary hypoplasia with poor development of the vasculature, with hyperplasia of the middle muscular layers and adventitia of pulmonary vessels. Hypoplasia and persistent pulmonary hypertension are the leading cause of high morbidity and mortality of this malformation. \(^{(14)}\)

The most common defect occurs on the left of the diaphragm (hernia Bochdalek 85%), posterolateral region but may be right in 15% of cases and bilateral in about 1-2%. The complete agenesis of the diaphragm and herniation are very rare events. \(^{(15)}\)

**Clinic:**
Most newborn with CDH have severe respiratory distress in the neonatal period, however, a lower percentage of posterolateral hernias and most parasternal remain asymptomatic beyond the newborn period and many of them will be researched as a radiological finding, even at older ages. \(^{(14)}\)

**Imaging studies:**
The sonographic features vary depending on which side of the hernia, making it easier to diagnose the HDC HDC left that right, because in the latter the echogenicity of the lung and liver are similar and gastric camera is intraabdominal. \(^{(15)}\)

Detection Ultrasonography (US) obstetrical can be performed from around the 18th week of pregnancy, depending on the time the content intraabdominal herniation into the chest occursutero. However, after 24 weeks, you can be performed more accurately with a sensitivity of 18-87%. Typical findings are gastric bubble or hepatic lobe at the level of the heart, bowel loops in the chest, dug abdomen, gallbladder and umbilical vein in abnormal position in the abdomen. \(^{(16)}\)

One of the utilities of prenatal US is the measurement of the relationship between the lung and head circumference, which is the product of orthogonal diameter of the right level earpiece plane heart divided by the head circumference in millimeters lung. This relationship was developed as a measure of pulmonary hypoplasia in the HDC on the left side, trying to correlate with survival. A lower ratio of 1 is associated with 100% mortality and a higher ratio of 1.4 with a survival rate of 100%. However, a considerable number of fetuses has intermediate values and the survival rate can range from 38-61%. \(^{(17-18)}\)

Magnetic resonance imaging (MRI) fetal provides additional information to the US by 38 to 50% and its greatest contribution is the measurement of lung volume. When the measurement of fetal lung volume is less than 15 to 25% of normal for gestational age, it has a mortality risk of 19 to 40% and if a lung is not visible, 18 to 62%. \(^{(19)}\)

In the postnatal stage, Rx thorax and abdomen is the diagnostic imaging method of choice. Initially shows a lack of air in the abdomen and thorax opacity committed by abdominal contents herniated airless, with mass effect and displacement of the mediastinal structures, as in our patient. \(^{(19)}\)

If the diagnosis is unclear HDC, you can enter a nasogastric tube and obtain a chest radiograph evaluating its distal end, to be found in the chest if there is herniation of the stomach. It may also be useful to inject a small volume of air through the probe, which will further facilitate diagnosis, without using contrasting means. Observe the course of umbilical catheters is also helpful. \(^{(20)}\)

**Forecast:**
The mortality of patients with CDH has been decreasing over time due to advances in prenatal diagnosis and adequate postnatal treatment planning methods, improving medical therapy before surgical repair. Thus, as in some centers survival incidence it reached 80%. 

**DIFFERENTIAL DIAGNOSTICS:**

Being the Cystic Adenomatoid Malformation, the congenital lobar emphysema and CDH are diseases that are part of unilateral hiperlúcido lung, the differential diagnosis must be made with all entities that are manifested in imaging techniques such as injuries arising signs of Entrapment. air accompanied with pulmonary lucidity radio areas that can get them or not to cause mediastinal shift, support of clinical data to be essential for proper diagnosis the main entities that should be discarded are:

- Pneumothorax
- pulmonary sequestration
- bronchogenic cyst

Of these three entities we only focus on the pneumothorax since in the last two pathologies although the clinic is similar and can make us suspect a hyperlucent down this is ruled out by imaging methods because they do not originate radio areas lucidity but most while dense images originate radio.

**PNEUMOTHORAX**

Is the presence of air in the pleural cavity with consequent lung collapse.  

**Incidence:**  
Spontaneous pneumothorax occurs in 1-2% of all newborn, only 0.5% and n is symptomatic. 

**Etiology:**  
Usually no history of respiratory distress, difficult delivery or suction of blood, mucus or meconium.  
Children who have undergone endotracheal intubation, resuscitation, meconium aspiration, pneumonia or ventilatory therapy are at increased risk.  

15 to 20% of pneumothorax are bilateral, most are bilateral and predominantly right. The radiological study is essential to establish the diagnosis.  
As frequent causes in the neonatal period are:

- Spontaneous:
  - 02.01% infants in the first day of life
  - 10% are symptomatic
  - Immediate neonatal period: by high pressure transpulmonary (1st breaths)

- iatrogenic:
  - Surgical interventions
  - Injuries
  - Untimely resuscitation maneuvers
  - Mechanic ventilation
• cardiorespiratory disease
  • Secondary to lung disease
    • Hyaline membrane disease
    • Aspiration syndromes
    • pneumonias
    • TTN
    • atelectasis
    • Hypoplasia - pulmonary agenesis

**DIAGNOSIS**

**CLINICAL MANIFESTATIONS:**
Sudden dyspnoea, distress onset moderate or severe respiratory, cyanosis, apnea, decreased peripheral tissue perfusion. \(^{(21)}\)

**PHYSICAL EXAM:** gas interposition syndrome can \(^{(10)}\) appear subcutaneous emphysema, abolition of breath sounds, bloat and increased vocal vibrations, pleural friction (rare).

**Imaging tests:**

Rx: the line of the visceral pleura (lung edge display), hyperlucency and absence of vascular pattern is identified \(^{(11)}\).

TC: It is ideal for detecting small, hidden pneumothorax or tabicados in previous position lung, you can rule out associated diseases.

**Figure 6:**
A: Scheme Spontaneous pneumothorax in the presence of air is observed in the right pleural cavity.
B: Rx: Area left pulmonary hiperlucidez with Bronchovascular absence of plot, associated with left pulmonary atelectasis secondary to pneumothorax.

**COMPLICATIONS:**
Tension pneumothorax, bilateral pneumothorax, pneumomediastinum, subcutaneous emphysema, Hemo-pneumothorax: break adhesions, Pio pneumothorax, chronic pneumothorax \(^{(10)}\).
CONFLICT OF INTERESTS:

The authors declare no conflict of interest.

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REVISTA DE LA FEDERACION ECUATORIANA DE RADIOLOGIA. No 10 Sep 2015 pag 18-25

EDITOR REVISTA DE LA FEDERACION ECUATORIANA DE RADIOLOGIA

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