

CONGENITAL LOBAR HYPERINFLATION (CLH)



<u>Case #7</u>: A newborn girl presented with subcostal retractions at birth. Chest x-ray was done shown in Figure A. Supportive and antibiotic therapy were provided. Follow-up study after three days is in Figure B. What is your likely diagnosis?

Synonyms: Congenital Lobar Emphysema (CLE), Congenital Lobar Overinflation (CLO) or Infantile Lobar Emphysema (ILE)

This is a progressive overdistension of the alveoli associated with obstruction of a lobar bronchus due to intrinsic narrowing, extrinsic compression or bronchial cartilage defect. It involves the left upper lobe > middle lobe > right upper lobe > right or left lower lobe.

There are two types which are distinguished histologically based on the number of alveoli: The *classic* or *hypoalveolar type* has normal or fewer than expected number but markedly overdistended alveoli. The *polyalveolar type* has increased normally inflated air spaces.

CLH may first be detected on prenatal US which can demonstrate a hyperechoic, hyperexpanded lobe from increased trapped fetal fluid, and fetal MRI may show a T2-hyperintense, hyperexpanded lobe. Bronchial atresia however has similar imaging findings. Chest radiographs after birth initially show an opacity (*Figure A*) from fetal lung fluid retention. As the fluid is reabsorbed by the lymphatic system, the fluid is replaced with air becoming hyperlucent (*Figure B*). Follow-up studies show progressive hyperinflation exerting mass effect on the adjacent structures.

References:

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