DSPC and SP-B were isolated from TA. DSPC amounts, ^{2}H-palmitate and ^{13}C-leucine enrichments were measured by gas chromatography mass spectrometry.

Results: DSPC amount and kinetics have been measured in 17 newborns, and SP-B kinetics in 18. DSPC median (interquartile range) fractional synthesis rate (FSR) was 5.0 %/day (3.5-8.5) in CDH patients vs 6.1 %/day (5.8-7.0) in controls (p=0.31). DSPC secretion time (ST) was 10.7h (6.9-11.5) vs 7.9h (5.2-14.0) (p=0.65).

SP-B FSR was 33.4 %/day (28.4-42.9) vs 52.0 %/day (40.0-83.4) (p=0.03); SP-B peak time was 26h (24-32) vs 18h (15-21) (p=0.02), both significantly lower in the CDH group. SP-B ST was 6.1h (4.0-9.9) vs 2.7h (1.1-7.1) (p=0.24), and half life 7.5h (5.2-9.1) vs 11.1h (5.8-17.3) (p=0.33).

Conclusions: Decreased and delayed SP-B synthesis was found in newborns with CDH compared to controls while DSPC kinetics was similar in both groups, which may contribute to severe respiratory failure in these patients.

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MUTATIONS OF SURFACTANT-RELATED GENES SP-B, SP-C AND ABCA3 IN UNEXPLAINED ALEOLAR/INTERSTITIAL LUNG DISEASE IN NEWBORN AND INFANTS

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Aims: To determine the role of surfactant-related gene anomalies in neonatal/infantile interstitial lung disease.

Methods: 22 infants >32weeks/< 1year with unexplained progressive alveolar/interstitial lung disease. SP-B, SP-C and ABCA3 sequencing. Optical (OM) and electronical (EM) microscopy of biopsy/autopsy lung tissue. Surfactant proteins immunofluorescence.

Results: 13/22 infants (59.1%) had lamellar bodies (LB) ultrastructural abnormalities suggesting genetic surfactant deficiency. Eleven infants (50.0%) carried ABCA3 mutations.

-Five (22.7%) had bi-allelic mutations (2 homozygote frameshift, 3 double heterozygote missense), showing small, dense LB with 1-2 denser cores and no normal LB on EM.

-Five (22.7%) had a single mutation (four missense, one intron), showing a mix of small and normal LB plus multivesicular bodies (MVB) on EM. On immunofluorescence, one had cytoplasmic mislocation of TTF-1 despite normal TTF-1 gene sequence, one had altered cleavage of proSP-B into SP-B, and one had reduced/misplaced alveolar capillaries.

-One infant (4.5%) carried a heterozygote SP-C mutation, plus a synonymous ABCA3 mutation, showing large, empty LB lacking normal surfactant layers on EM.

-One infant (4.5%) with normal LB + MVB had a heterozygote SP-B mutation.

-One infant (4.5%) with small dense LB (similar to homozygote ABCA3 mutants) had no mutation identified.

In the 9 remaining infants (40.9%), one had alveolar proteinosis, one had COX-deficiency, one had Niemann-Pick C disease, two had idiopathic pulmonary fibrosis, four remained cryptogenetic.

Conclusions: ABCA3 mutations are the most frequent abnormality. Although EM is indicative in most (92%) cases, molecular mechanisms are elusive in >50% of cases, likely involving complex, multiple surfactant-related gene interactions.

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ROLE OF CORD STRESS HORMONE LEVELS FOR DEVELOPING TRANSIENT TACHYPNEA OF NEWBORN IN CESAREAN DELIVERED LATE PRETERM AND TERM INFANTS

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Background and aims: Failure of adequate and timely clearance of fetal lung fluid results in transient tachypnea of newborn (TTN). Of the several factors that have been proposed to effect sodium reabsorption some have been investigated including glucocorticoids, oxygen, catecholamines, thyroid hormones, surfactant. The aim of this study was to investigate the role of cord stress hormones;