

MULTIMODAL IMAGING IN THE CONGENITAL PULMONARY LYMPHANGIECTASIA-CONGENITAL CHYLOTHORAX-HYDROPS FETALIS CONTINUUM

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ABSTRACT

We report on three infants with congenital chylothorax (CC) and congenital pulmonary lymphangiectasia (CPL). CPL appears to be a characteristic pathological finding in CC. Through the use of lymphoscintigraphy and computed tomography, this study suggests that CC and CPL are strongly correlated entities and that the dysplasia of the lymphatic system results in a pulmonary lymphatic obstruction sequence. The initial microscopic dilatation of the lymph channels may lead to progressive weeping of lymphatics and, consequently, to pleural effusion. Non-Immune Hydrops Fetalis (NIHF) may be the final consequence of impaired systemic venous return and may help to explain pleural-pulmonary involvement in this generalized lymph-vessel malformation syndrome.