Opis choroby *

Definicja

A rare respiratory tumor characterized by an aggressive, malignant, dysontogenetic neoplasm of intrathoracic (pulmonary, pleural, or combined) mesenchyme occurring in young children. Three subtypes can be distinguished, type 1 being purely cystic, type 2 cystic and solid, and type 3 purely solid. Type 1 lesions may progress to the more malignant types 2 and 3, which are associated with central nervous system and bone metastasis. The tumor is often part of pleuropulmonary blastoma family tumor and dysplasia syndrome. It can also be associated with multilocular cystic nephroma or other neoplasms. Patients usually present with dyspnea or other respiratory problems, and sometimes pneumothorax.

Dane

Klasyfikacja
Choroba

Kod ORPHA
64742

Kod OMIM
601200

Kod ICD10
C34.1

Kod ICD11
2C25.Y

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*Źródło

orphanet