A rare lung disease and its association with Congenital Heart Disease

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Pulmonary Interstitial Glycogenosis (PIG) is a rare condition of undetermined incidence of interstitial lung disease, with unique histologic features. The cellular and molecular mechanisms of this disease are not fully elucidated. The clinical findings of respiratory distress and hypoxemia are among the clinical features which can occur among infants with congenital heart disease (CHD).

Methods: Electronic medical records (EMR) from the dates 8/1/2009 to 5/5/2016, using ICD (international classification of disease) codes corresponding to (i) Transposition of the Great Arteries (TGA) (ii) Tetralogy of Fallot (TOF) (iii) persistent pulmonary hypertension (PPH), and (iv) pulmonary interstitial glycogenosis (PIG), were searched to identify cases of associated PIG and CHD or PPH. A literature search using PubMed/Medline from 2002 to 2016 was conducted to identify cases of CHD and PIG, and PPH and PIG.

Results: The EMR search identified two cases of PIG, but none were associated with TGA or TOF. One of the two cases were associated with PPH. The literature search identified eight cases of a PIG in association with CHD or PPH as described in seven articles.

Conclusions: There is an association between PIG and cardiovascular malformations. Overlapping molecular pathways involved in the developing cardiovascular system and mesenchyme of the lung should be further studied. Among particular pathways suggested as the topic of future study include NOTCH and VEGF signaling pathways.

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