

シンポジウム | 肺循環

シンポジウム02 (I-S02)

肺循環「6th World Symposium on PH 2018を、どう解釈し、どう活かすべきか？」

座長:土井 庄三郎 (国立病院機構 災害医療センター)

座長:中山 智孝 (高知大学医学部 小児思春期医学)

2020年11月22日(日) 15:30 ~ 18:00 Track2

[I-S02-7]小児肺高血圧症(PH)の診断と治療：欧州小児肺血管疾患ネットワーク(EPPVDN)合意声明2019年改訂版

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キーワード：肺高血圧症, 気管支肺異形成, 新生児遷延性肺高血圧症

Aim: To define and develop effective, innovative diagnostic methods and treatment options in all forms of pediatric pulmonary hypertensive vascular disease.

Methods: Clinical trials, meta-analyses, guidelines, and other articles that include pediatric data were searched using the term "pulmonary hypertension" and other keywords. Class of recommendation and level of evidence were assigned based on ESC/AHA definitions and on pediatric data only, or on adult studies that included >10% children or studies that enrolled adults with CHD. New definitions presented by the World Symposium on PH 2018 were included.

Results: We generated 10 tables with graded recommendations. The topics include diagnosis, monitoring, genetics, biomarkers, cardiac catheterization, echocardiography, cardiac MR/chest CT, associated forms of PH, ICU, lung transplantation, and treatment of pediatric PH. A simplified adult PAH risk score based on the 2015 ESC/ERS PH guidelines emphasizes particularly the prognostic value of mean right atrial pressure, cardiac index, WHO functional class, and NT-proBNP. We subsequently developed a pediatric PAH risk score that needs to be validated in future studies. The evolving strategy of upfront (or early rapid sequence) combination pharmacotherapy may further improve outcome of pediatric PAH. A set of specific recommendations on the management of PH in low-income regions was developed.

Conclusions: These executive, up-to-date guidelines provide a specific, comprehensive, detailed but practical framework for the optimal clinical care of children and young adults with PH.