Advances in targeted therapy for pulmonary arterial hypertension in children

Li Li[1], Xinyu Zhu[1], Xiaojie Chen[1], Jieyun Gao[1], Chunchun Ding[1], Mengrou Xu[1], Min Zhang[1], and Shixun Ma[1]

[1]Gansu Provincial Hospital

July 5, 2022

Abstract

Pulmonary arterial hypertension (PAH) is a rare and devastating disease of the pulmonary vasculature with a high morbidity and mortality rate in infants and children. Currently, treatment approaches are mostly based on adult guidelines and pediatrician clinical experience, focusing on specific pulmonary antihypertensive therapy and conventional supportive care. The advent of targeted drugs has led to significant advances in the treatment of PAH in children, including endothelin receptor antagonists, phosphodiesterase type 5 inhibitors, and prostacyclins, which have been studied and proven to improve hemodynamics and functional class in children PAH. A new targeted drug, riociguat, is assessing its safety and efficacy in clinical trials. However, more randomized controlled studies are needed to evaluate the combination of drugs, treatment strategies, and clinical endpoints of targeted therapy in children PAH. In this review, we summarize the research advances of PAH-targeted therapy in children over the last decade in order to provide a theoretical basis for future studies.

Hosted file

`manuscript.doc` available at https://authorea.com/users/493328/articles/575766-advances-in-targeted-therapy-for-pulmonary-arterial-hypertension-in-children