

Diagnosis and Management of Pulmonary Arterial Hypertension in the Pediatric Population (PH)

Hanaa Hasan Banjar, M.D, FRCPC*

Pulmonary arterial hypertension (PH) is a rare disease that affects either sex at any age. It is frequently under-diagnosed and many patients present with prolonged interval between onset of symptoms and diagnosis, which delays their treatment and shortens their prognosis yet further. Mortality associated with PH is extremely high. Once the diagnosis has been confirmed, mean survival among adults is 2.8 years and less than one year among children. Advances in the understanding of the mechanisms involved in the patho-biology of PH associated with other conditions have focused on molecular biology, developmental biology and genetics. Together with epidemiological and natural history studies, collaborative efforts between the scientific community and the industry have led to a surge in clinical trials over the past decade with six new medications which are approved by the FDA that lead to improvement in the median survival, Quality of life, 6 minutes walk test, functional class and delayed progression of the disease.

In this review, we discuss the recent diagnostic and treatment modalities which are appropriate to the pediatric population.

Bahrain Med Bull 2008; 30(4):