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Sildenafil: Experience in Children with or without Pulmonary Hypertension and Congenital Cardiac Defects

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Background: Data regarding dosage, safety, and clinical and hemodynamic responses to Sildenafil for management of patients with congenital heart disease (CHD) with or without pulmonary artery hypertension(PAH) has only been reported sporadically.
Aim: Describe the cohort of children in whom Sildenafil was used and determine the clinical and corresponding hemodynamic response.
Methods: We retrospectively studied 85 patients (108 records) who were treated between January 2008 and December 2010 (mean age 4.2years; 42females; mean weight 13.5kg). Patients were classified into 4 groups; 1: Idiopathic PAH(N=8); 2:Biventricular circulation with PAH, including Cardiomyopathies and CHD (N=30); 3:Right ventricular dysfunction (Fallot type and Heart transplantation, N=9); 4:Univentricular circulation (N=59). We recorded dosage, duration, side effects, and setting of treatment: Outpatient(N=4); Hospitalized(N=47); Post-heart surgery(N=56). The following objectives were analysed:1)Respiratory improvement (withdrawal of mechanical ventilation, supplementary oxygen or nitric-oxid);2)Echocardiographic improvment (PAH estimation and ventricular function);3)Clinical situation (oxygen saturation and functional status) ; and others, such as decrease of debit drains on starting, at 6 months or on terminating treatment. Hemodynamic response in those undergoing catheterization pre(N=85) and post (N=65) treatment was analysed.
Results: The mean initial dosage was 1.68mg/kg/d (Range 0.4-4) and mean duration of treatment was 12.6 months. Escalating to the maximum dosage occurred within 48 hours in 65.4% (mean maximum dosage 2.36). Side effects were reported in 17 patients, requiring withdrawal in 2 (1.9%) both postop. An improvement was observed in at least one objective on starting in 66 records(70%), especially in respiratory evolution 42.4%. Improvement in pulmonary vascular resistance and mean pulmonary pressures was observed in Groups 1, 2 and 4 ($P<0.05$). Improvement in flow ratio (Qp/Qs) was observed in Group 3 ($P<0.05$). The children with echocardiographic ventricular dysfunction pretreatment(N=28) had a worse initial response ($P=0.006$), except for patients in Group 4 (acute right ventricular dysfunction posttransplant, N=4) which improved.
Conclusion: Oral administration of Sildenafil in children with or without PAH and CHD is safe and has a favourable clinical and hemodynamic response with the use of standard dosage, except in those patients with worse ventricular function.