Liječenje plućne hipertenzije specifičnim vazodilatatorima u bolesnika s Eisenmengerovim sindromom – naša iskustva

Vasodilatory treatment of pulmonary hypertension in patients with Eisenmenger's syndrome: our experience

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UVOD: Eisenmengerov sindrom je najrazvijeniji oblik plućne arterijske hipertenzije uzrokovane kongenitalnim srčanim greškama. Tri su skupine lijekova do sada odobrene za liječenje endotelne disfunkcije plućnog krvožilja u ovih bolesnika: prostanoidi, antagonisti endotelinskih receptora i inhibitori fosfodiesteraze-5.

METODE: Retrospektivna analiza podataka iz Registra plućne hipertenzije Klinike za plućne bolesti Kliničkog bolničkog centra Zagreb. Od ukupno 33 pacijenta sa korigiranim ili nekorigiranim srčanom greškom u Registru koji prati pacijente od 2000. do 2014. god. njih 18 je liječeno specifičnim vazodilatatorima. Analizirani su demografski podatci te varijable koje utječu na procjenu funkcionalnog statusa pacijenta (NYHA status, šestominutni test hoda i saturacija arterijske krvi kisikom) prije početka liječenja i na posljednjoj kontroli.

REZULTATI: Od rujna 2000. do rujna 2014. god. liječeno je 18 pacijenata s verificiranim Eisenmengerovim sindromom. Svi pacijenti stariji su od 18 godina i 13/18 (72 %) su ženskog spola. Prosječna dob u vrijeme početka liječenja specifičnim vazodilatatorima je 34,3 godine, a trajanje istog u prosjeku je 4,4 godine. Pacijenti su liječeni bosentanom (11 pacijenata), sildenafilom (12 pacijenata), iloprostom (6 pacijenata) te kombinacijama navedenih, a jedan pacijent liječen je ambrisentanom. U ovih 18 pacijenata uključeno je 10 pacijenata sa VSD-om (od kojih jedan korigirani), četiri s ASD-om (1 korigirani), jedan pacijent s transpozicijom velikih krvnih žila, dva pacijenta s arteriovenskim kanalom i Downovim sindromom te jedan pacijent s aortopulmonalnim prozorom. Funkcijski status pacijenata značajno se poboljšao nakon uključivanja lijeka (NYHA 2,2±0,5 vs. NYHA 2,8±0,7, P=0.032). Saturacija arterijske krvi prije i nakon uvođenja lijekova nije se značajno mijenjala (satO₂ 87±8 vs. 87±9%, P=0.941), kao ni nalaz šestminutnog testa hoda (hodna pruga 323±93 vs. 353±69 m, P=0.532).

ZAKLJUČAK: Liječenje plućne hipertenzije specifičnim vazodilatatorima u skupini naših pacijenata ukazuje na dugoročni benefit s obzirom na poboljšanje funkcijskog statusa pacijenata. Nije uočen pad saturacije kisikom u ispitivanoj populaciji bolesnika.

INTRODUCTION: Pulmonary arterial hypertension in patients with congenital heart disease often results in Eisenmenger's syndrome. There are three major drug classes approved for treatment of endothelial dysfunction of pulmonary vasculature: prostanoids, endothelin receptor antagonists and phosphodiesterase-5 inhibitors.

METHODS: A retrospective review of the pulmonary hypertension Registry of the Clinic for Pulmonary Diseases University Hospital Centre Zagreb was performed. We analyzed the data from 18 GUCH (grown-up congenital heart disease) patients treated with specific vasodilators from September 2000 to September 2014. Demographic parameters, functional class, six minute walk test and oxygen saturation were assessed before and during the treatment.

RESULTS: From September 2000 to September 2014, 18 patients diagnosed with Eisenmenger's syndrome were treated with pulmonary vasodilators. Patients were older than 18 years of age and 72% of them were female. Their mean age was 34.3 years. The average duration of treatment was 4.4 years. 11 were treated with bosentan, 12 were taking sildenafil, 6 patients were treated with iloprost and some were taking combinations of the aforementioned drugs. One patient was treated with ambrisentan. Eisenmenger's syndrome was caused by VSD in 10 patients (1 underwent operative VSD closure), ASD in 4 patients (1 was corrected), transposition of the great arteries in 1 patient, AV canal in the setting of Down's syndrome in 2 patients and AP window in one patient. The functional status improved after treatment (NYHA 2.2±0.5 vs. NYHA 2.8±0.7, P=0.032). The arterial saturation was not adversely affected by the treatment (87±8 vs. 87±9%, P=0.941). The observed improvement in the 6MWT did not reach statistical significance (323±93 vs. 353±69 m, P=0.532).

CONCLUSIONS: Modulation of the pulmonary vascular tone may improve the functional status of GUCH patients complicated with Eisenmenger's syndrome. Specific pulmonary vasodilatory treatment did not adversely affect oxygenation in the studied cohort.

LITERATURE

1. D'Alto M, Diller GP. Pulmonary hypertension in adults with congenital heart disease and Eisenmenger syndrome: current advanced management strategies. Heart. 2014;100(17):1322-8.

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