Kongenitalni totalni atrioventrikularni blok u odrasle osobe

Congenital complete atrioventricular block in an adult

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Uvod: Totalni atrioventrikularni blok (TAVB) je najčešći oblik poremećaja atrioventrikularnog provođenja, a javlja se u 1/15 000 novorođenčadi. Može se javiti kao posljedica strukturne bolesti srca ili kao izolirani poremećaj. Procijenjena stopa mortaliteta među odraslima s izoliranim kongenitalnim TAVB iznosi 5%. Točan mehanizam nastanka izoliranog kongenitalnog TAVB je nepoznat. Pretpostavlja se kako imunosni odgovor igra glavnu ulogu, nastao uslijed transplacentalnog prijelaza majčinih autoantitijela na antigene jezgre, napose SSA/Ro i SSB/La. Upalni odgovor vodi oštećenju tkiva i fibrozi provodnog sustava srca. Ostali uzroci uključuju virusnu infekciju i produljeni QT sindrom. Kako su bolesnici uglavnom asimptomatski, dijagnoza se obično postavlja rutinskim 12-kanalnim EKG zapisom ili 24-satnim elektrokardiografskim zapisom (Holter EKG), testom opterećenja i ehokardiografijom.[1,2]

Prikaz slučaja: 22-godišnja bolesnica je zaprimljena u bolnicu radi vrtoglavica. Nekoliko mjeseci ranije proširena neurološka i otorinolaringološka obrada je isključila postojanje neurološke i vestibularne patologije. 24-satnim Holter EKG snimanjem verificirana je atrioventrikularna disocijacija, s prosječnim odgovorom klijetki od 47/min (interval 32-88/min). Test opterećenja je bio uredan, s adekvatnim kronotropnim odgovorom, maksimalno do 158/min. U početku testa prisutan je atrioventrikularni blok (AVB) 2:1, dok na vrhuncu opterećenja se registrira AVB-a tip I. Tilt-up table test je isključio ortostatsku hipotenziju i vazovagalnu komponentu. Ehokardiografija je pokazala minimalni prolaps prednjeg mitralnog kuspisa, blagi stupanj mitralne i trikuspidalne regurgitacije. Ponovljeni Holter EKG je pokazao sinusni ritam, s prosječnim odgovorom klijetki od 54/min (interval 32-114/min) te intermitentni AVB drugog stupnja Mobitz I i TAVB. Dodatno se učini test opterećenja po preskakućem Bruce protokolu, u tijeku kojeg bolesnica razvije pad frekvencije srca koji jasno korelira sa presinkopalnim atakama. Postavi se indikacija za ugradnjom trajnog ES koji je implantiran s izravnom stimulacijom Hisovog snopa kako bi se izbjegla indukcija disinkronije.

Zaključak: Kod osobe s kongenitalnim totalnim atrioventrikularnim blokom bez strukturne bolesti uz demarkaciju simptoma testom opterećenja prema Bruce preskakućem protokolu postavili smo indikaciju za ugradnjom trajnog ES.

Introduction: Complete congenital atrioventricular block (CCB) is the most common type of atrioventricular conduction impairment with the incidence of 1 in 15 000 births. It may occur as a result of a structural heart disease or it can be isolated. Estimated mortality rate among adults with isolated CCB is 5%. The exact mechanisms of isolated CCB occurrence is still unknown. The assumption is that the immune response has a major role in its emergence due to transplacental passage of maternal autoantibodies to the nuclear antigens, predominantly SSA/Ro and SSB/La. Consequent inflammation leads to injury and fibrosis of the conduction heart system. Other possible causes include viral infections and long QT syndrome. As patients are predominantly asymptomatic, routine 12-lead ECG is often the first tool in making a diagnosis of CCB. Other diagnostic procedures are 24-hour electrocardiographic recordings (Holter ECG), exercise stress test, and echocardiography.[1,2]

Case report: 22-year old female patient was admitted to hospital due to dizziness. Few months earlier, extensive neurological and otorhinolaryngological examination was done, which showed no signs of any central nervous system or vestibular pathology. 24-hour Holter ECG verified atrioventricular dissociation, with average heart rate 47 (interval 32-88) beats per minute (bpm). Exercise stress test was normal, with adequate chronotropic response, maximum to 158 bpm. In the beginning of the test 2:1 atrioventricular block (AVB) was observed, while in the peak load AVB type I was noticed. Tilt-up table test excluded an orthostatic and vasovagal component. Echocardiography showed minimal prolapse of the mitral anterior cusp with mild mitral and tricuspid regurgitation. Repeated Holter ECG showed sinus rhythm, with average heart rate of 54 bpm (interval 32-114 bpm), and intermittent second degree AVB, Mobitz I and total AVB. Additional testing was performed using overlapping Bruce protocol during which significant decrease in heart rate was registered and followed by presyncopal episodes. Therefore permanent pacemaker was implanted which stimulated the His bundle in order to avoid dyssynchrony.

Conclusion: In patient with CCB, without structural heart disease, using overlapping Bruce protocol we have unmasked presyncopal symptoms, and therefore made an indication for permanent pacemaker implantation.

LITERATURE

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