Brugada sindrom – prvi rezultati ajmalinskog testiranja

Brugada syndrome – first results of ajmaline testing

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Uvod: Brugada sindrom (BS) je genetska bolest karakterizirana tipičnim promjenama u EKG-u i povišenim rizikom od nagle srčane smrti (NSS). Blokatori natrijskih kanala mogu demaskirati bolest induciranjem tipičnog Brugada tip I uzorka u EKG-u[1,2]. Do nedavno ajmalin nije bio dostupan u RH te nisu sistematski provođena ajmalinska testiranja.

Metode: Od prosinca 2015. ajmalin je ponovno dostupan u Kliničkom bolničkom centru Zagreb. Ajmalinski test bio je indiciran u pojedinaca s obiteljskom anamnezom NSS ili nerazjašnjene sinkope uz Brugada tip 2 obrazac u EKG-u, odnosno u članova obitelji otkrivenog probanda. Testiranje je bilo provođeno u elektrofiziološkom (EP) laboratoriju koristeći „EP Medsystems" uređaj. Korištene su standardne EKG postavke, (25 mm/s, 1 mV/1 cm). Osim standardnih odvoda snimani su V1 i V2 odvodi u trećem interkostalnom prostoru. Ajmalin je primljenjivan dozi od 1 mg/kg unutar 5 minuta. Nakon snimljenog bazičnog EKG-a, sukcesivno su snimani zapisi na 0,7 mg/kg (3 min aplikacije), 1 mg/kg (5 min aplikacije) te faza ispiranja (2 min nakon aplikacije). Pozitivnost testa bila je definirana kao tipična tip I morfologija u V1 ili V2 odvodima sa ST elevacijom od ≥ 2 mm u trećem ili četvrtom interkostalnom prostoru[3]. U slučaju pozitivnog testa učinjena je EP studija.

Rezultati: Testirano je ukupno 14 bolesnika (7 muškaraca, prosječna dob 35,9 ± 15,7 godina). Pet od 14 testiranja bilo je pozitivno (35,7 %). Otkrivena su 4 BS probanda te jedan član obitelji kod kojeg je proband bio je poznat od ranije. Osam (57,1 %) pojedinaca testirano je u sklopu obiteljske anamneze BS. Kod jednog od 5 bolesnika (20 %) EP studija je bila pozitivna, a kod 3 od 5 (60 %) BS pozitivna bolesnika ugrađen je ugradbeni kardioverter defibrilator (ICD). Uz tip I obrazac ajmalinskog testa, pozitivna EP studija ili nerazjašnjena sinkopa indicirale su ugradnju ICD-a. Jedan bolesnik (33 %) imao je uključivanje ICD-a koje je bilo neadekvatno, uslijed do tada nepoznate fibrilacije atrija s brzim odgovorom klijetki.

Zaključak: Brugada sindrom važan je uzrok NSS u bolesnika sa strukturno zdravim srcem. Ajmalinski test omogućava otkrivanje ove bolesti te prevenciju NSS u inače mladih zdravih pojedinaca. Osim izbjegavanja određenih lijekova, ugradnja ICD-a ključan je dio liječenja ovog sindroma.

Introduction: Brugada syndrome (BS) is a genetic disease characterized by typical changes in the ECG and an increased risk of sudden cardiac death (SCD). Sodium channel blockers may unmask disease by inducing a typical Brugada type I pattern in the ECG[1,2]. Until recently, ajmaline was not available in the Republic of Croatia and ajmaline testing was not routinely conducted.

Methods: From December 2015 ajmaline is available at University Hospital Centre Zagreb. Ajmalin test was indicated for individuals with a family history of SCD or unexplained syncope with type 2 Brugada pattern, or the family members of discovered proband. Testing was conducted in electrophysiologic (EP) lab using "EP Medsystems" hardware. Standard ECG settings were applied (25 mm/s, 1 mV/1 cm). In addition to standard precordial leads, V1 and V2 were recorded in the third intercostal space. Ajmaline was applied in a dose of 1 mg/kg within 5 minutes. After recording the basic ECG, traces were recorded at 0.7 mg/kg (3 min), 1 mg/kg (5 min) and washout (2 min after the administration). Test was defined as positive when a typical type I morphology in V1 or V2 leads with ST elevation of ≥2 mm occurred[3]. In case of a positive test EP study was performed.

Results: The study included 14 patients (7 men, mean age 35.9±15.7 years). Five of the 14 tests were positive (35.7%). Four BS probands were discovered, and one family member tested positive. Eight (57.1%) individuals were tested as family screening. In one of the 5 ajmaline positive patients (20%) EP study was positive, while 3 out of 5 (60%) patients positive BS received an implantable cardioverter defibrillator (ICD). Unexplained syncope or positive EP study alongside with positive ajmaline test indicated ICD implantation. One patient (33%) received inadequate ICD shock, due to a previously unrecognized atrial fibrillation with a rapid ventricular response.

Conclusion: Brugada syndrome is an important cause of sudden cardiac death in patients with structurally normal heart. Ajmalin test allows detection of the disease and prevention of SCD in otherwise young healthy individuals. In addition to avoiding certain drugs, implantation of an ICD is a key part of the treatment of this syndrome.

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